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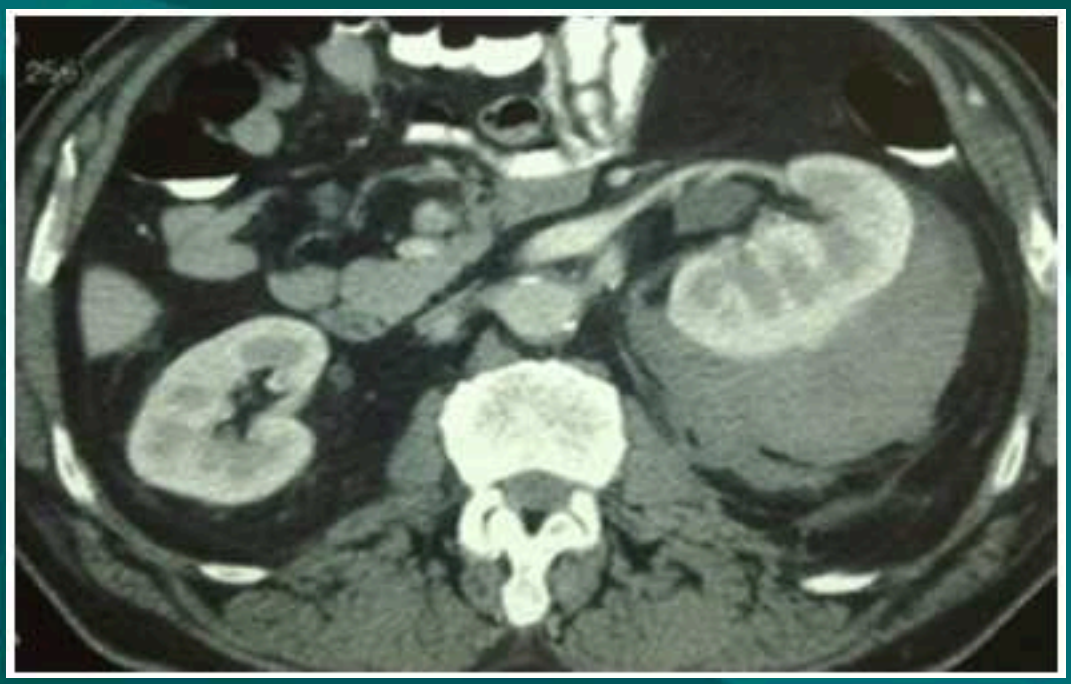

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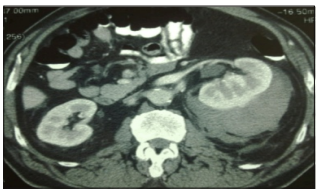

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CASE SERIES OPEN ACCESS

Management of millers class III marginal tissue recession associated with endodontic lesion: Report of two cases managed using second-stage surgery

Sangeeta Singh

ABSTRACT

Introduction: Endo perio lesions are common conditions that are often difficult to diagnose and are persistent if not treated completely. However, if the patient's history is taken carefully and thorough evaluation of all possible routes of infection is carried out, a properly done endodontic treatment is sufficient to eliminate the infection. However, wherever a secondary periodontal involvement exists, it requires specific therapy to achieve success. Case Series: In the first case, the involved maxillary left first premolar had a severe marginal tissue recession completely exposing the buccal root. The case was further complicated by the presence of an endodontic lesion. After successfully completing endodontic therapy, a free gingival autograft was placed to increase the zone of attached gingiva. Subsequently, a connective tissue graft was placed using pouch and tunnel technique to augment the zone further. The second case had a Millers class III recession associated with an endo perio lesion. This case was managed by using a resorbable membrane with a bone graft substitute to correct the osseous defect in the first stage after a successful endodontic therapy. The second-stage surgery was done using an envelope technique for connective tissue grafting. In both the cases, there was an increase in width and thickness of the zone of attached gingiva following the two-step surgical procedure after a successful endodontic therapy. Conclusion: Successful treatment reported in both the cases can be attributed to a correct diagnosis, successful endodontic therapy and an increased zone of attached gingiva achieved using a two-step surgical technique.

Keywords: Marginal tissue recession, Endo perio lesions, Root coverage, Pouch, Tunnel

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INTRODUCTION

The relationship between periodontal and pulpal disease was first described by Simring et al. in 1964 [1]. Since then, the term 'endo perio lesion' has been used to describe lesions due to inflammatory products found in varying degrees in both periodontium and pulpal tissues. In most cases of endo perio lesions, clinical symptoms disappear following successful endodontic therapy. However, it becomes essential to correct the periodontal defect simultaneously in these cases to prevent recurrence, and to improve the functional status of the tooth [2]. Some of the most important functional goals in the treatment of mucogingival problems are arresting the progression of gingival recession and

improving the ability for plaque control in cases with healthy and diseased marginal tissues. Marginal tissue recession as a clinical entity has been documented since the last century. The earlier concept of a defined width of attached gingiva necessary to maintain oral hygiene does not hold true now [3]. It is essential to carry out root coverage surgery whenever concerns such as aesthetics, sensitivity, susceptibility to root caries, pulpal symptoms due to exposure of root, food lodgment and plaque deposition exist. Currently, accepted procedures for root coverage include coronally advanced flap, free mucosal graft, sub epithelial connective tissue graft, guided tissue regeneration and acellular dermal matrix.

CASE SERIES

Case 1: A 37-year-old male patient reported with the complaints of sensitivity, foul smell and pus discharge from the maxillary left first premolar since one month. The tooth was previously sensitive to hot and cold, and had recently developed spontaneous pain and pus discharge. He had visited various dental centers during past eight years but no definitive treatment had been done. Clinically, there was no attached gingiva in the region of 24 and the buccal root was completely exposed. An intraoral radiograph showed a periapical radiolucency (Figure 1A–C). A diagnosis of Millers class III marginal tissue recession (MTR) associated with an endodontic lesion in the maxillary left first premolar was made. After completing endodontic therapy, the first step to correct the mucogingival defect was to increase the zone of attached gingiva using a free gingival autograft from the palate (Figure 2A–D). Once the graft was successful and stable, after one month the second surgery was carried out utilizing a connective tissue graft from the palate which was placed using the pouch and tunnel technique [3] (Figure 3A–C).

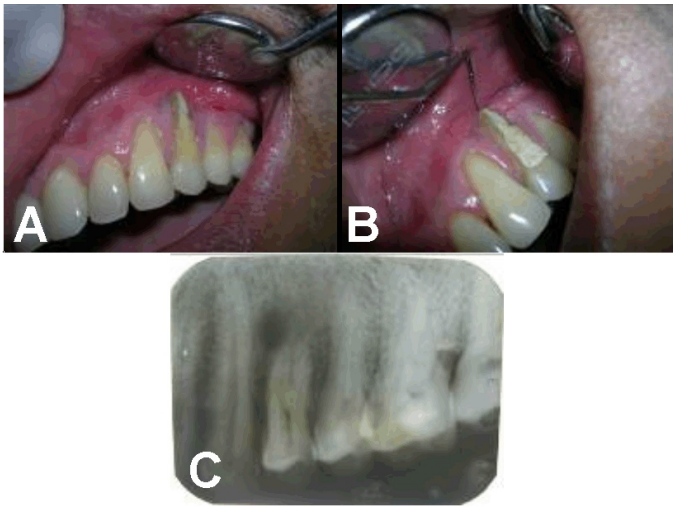

Figure 1: (A) Millers class III MTR in 24, (B) Complete exposure of buccal rootbeyond apex, (C) Periapical radiolucency in relation to 24.

Case 2: A 26-year-old male patient reported with complaints of pus discharge and pain in relation to maxillary left first premolar since two months. His history revealed that he had pain in the tooth since one year but was intermittently taking some medicines from the local physician. The pain would subside for some time and recur again. The pus discharge had started two months back with no relief despite taking medicines. Clinically, the area appeared inflamed. There was tenderness on probing and there was a pocket of > 5 mm on buccal, mesial and distal aspect. Radiograph revealed a periapical radiolucency and an infrabony defect on distal aspect of 24 (Figure 4A–B). This case was diagnosed as Millers class III MTR associated with a periapical lesion in 24. After a successful endodontic therapy, first surgery was carried out to correct the intraosseous defect. After debridement there was an osseous defect along the buccal root. A bone graft substitute along with a resorbable membrane was placed

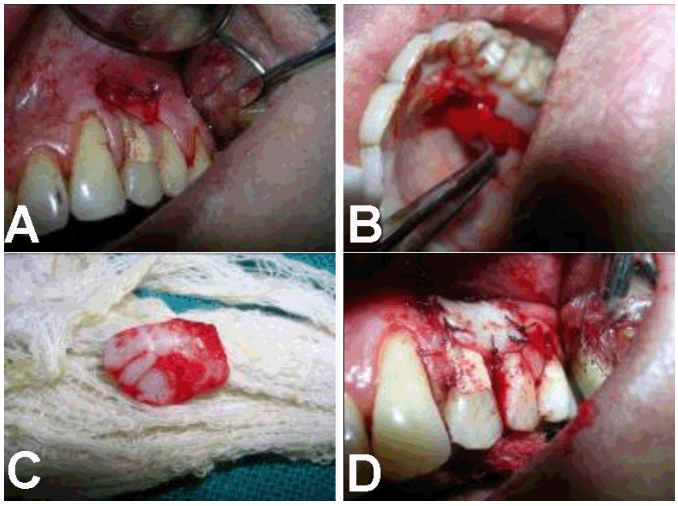

Figure 2: (A) Recipient site prepared, (B) Free autogenous graft being harvested from palate, (C) The graft, (D) Graft sutured.

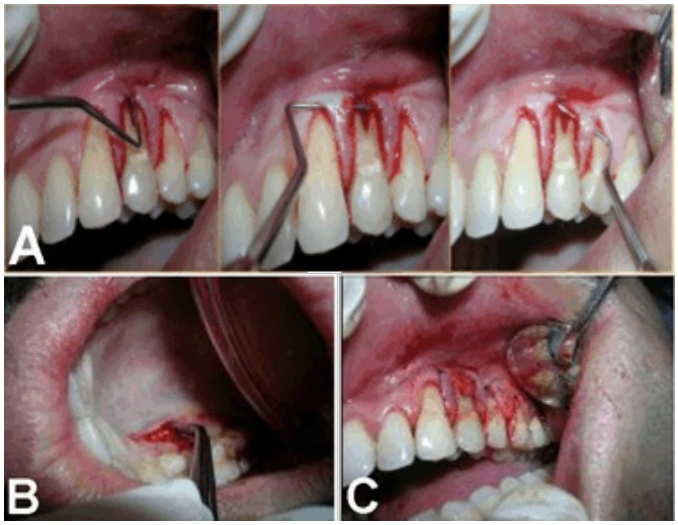

Figure 3: (A) Tunnel prepared for the connective tissue graft, (B) Graft being harvested from the palate, (C) Graft sutured.

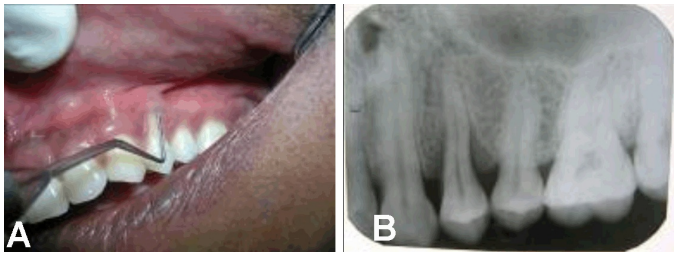

Figure 4: (A) Millers Class III recession of 24, (B) Radiograph showing periapical lesion and infrabony defect at distal aspect.

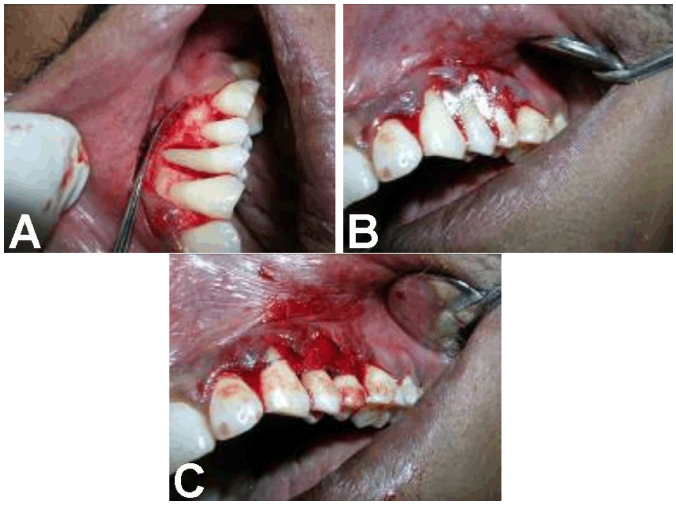

Figure 5: (A) Osseous defect surrounding buccal root, (B) Bone graft substitute placed, (C) Resorbable membrane sutured.

to cover the defect. The flap was then sutured (Figure 5A–C). At the time of review, two months later the tooth appeared healthy. However, the patient was unable to keep the tooth completely plaque free due to the recession. Therefore, a second surgery was carried out to place a connective tissue graft using the envelope technique to achieve root coverage (Figure 6A–E). In both the cases after the primary lesion was treated, the two-step surgery helped in not only creating a zone of attached gingiva but also enhancing the thickness of the attached gingiva (Figure 7A–B).

DISCUSSION

The pulp and the periodontium share a interrelationship via the apical foramen, lateral canals and dentinal tubules [1]. The inflammatory by products from the pulp can leach out and trigger an inflammatory response in the periodontium [4]. Both the cases discussed here had a primary endodontic lesion with secondary periodontal involvement [5]. The involvement was severe enough to have exposed the buccal root completely in the first case. The goal was to treat the primary infection as well as achieve functional restoration of the periodontium rather than esthetics [6].

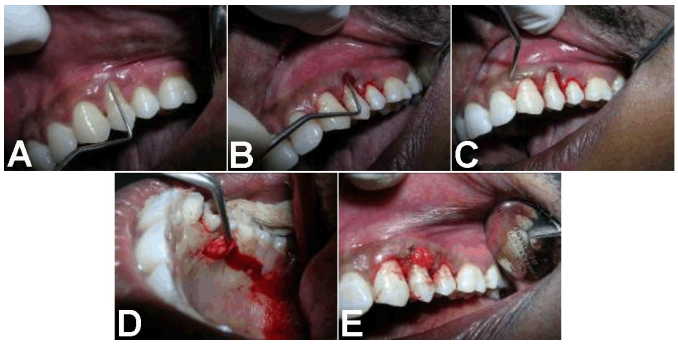

Figure 6: (A) A vertical recession of 3 mm on 24, (B) Undermining the buccal aspect, (C) Creating an envelope, (D) Harvesting connective tissue graft, (E) Graft sutured.

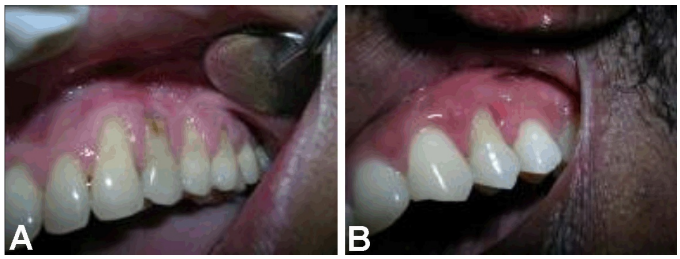

Figure 7: (A) Case 1 postoperative nine months, (B) Case 2 postoperative six months.

Marginal tissue recession requires treatment for many reasons such as impaired aesthetic appearance, root sensitivity, cervical caries or abrasion. Two surgical techniques have been described that use free gingival graft for root coverage. The technique proposed by Bernimoullin et al. involves two surgical steps. The first step consists of creating attached gingiva by means of free gingival graft and second step involved coronal positioning of grafted tissue to cover the gingival recession. This indirect technique has advantages over other techniques because it ensures development of an adequate band of attached gingival [7]. The first case had a severe marginal tissue recession complicated by the presence of a long standing endodontic lesion. After successfully completing endodontic therapy, the first step was to place a free gingival graft from the palate at the recession site. This was important to create a zone of attached gingiva where none existed. Free gingival graft was the technique of choice because it has been documented as the most predictable method to increase the apico-coronal dimension of the keratinized mucosa despite the advent of subepithelial connective tissue graft and allogenous grafts like AlloDerm [8]. Once the graft was successful, after four weeks a connective tissue graft from the palate was procured and placed using the pouch and tunnel technique to increase the thickness as well as achieve some root coverage in region of tooth 24. The use of tunnel procedure preserves the interdental papilla and this facilitates an early and accelerated initial wound healing. The tunneling also applies less traction and preserves the gingival height [9]. The elimination of vertical incision, which is used in

subepithelial connective tissue grafting, ensures complete coverage of the connective tissue graft by the flap thus aids in faster healing as well as excellent color matching. The pouch and tunnel procedure may be of advantage as compared to coronal repositioned flap since there is minimum trauma to the recipient site and there is predictable root coverage [10]. The second case was treated using the same approach, i.e. treating the primary lesion first and later correcting the osseous defect using a resorbable membrane with bone graft substitute. A second surgery done using connective tissue graft from the palate and placing it using an envelope technique helped in further augmenting the attached gingiva, thus ensuring long term stability of results.

The cases discussed in this case series were challenging not only because of the presence of an endo perio lesion but there was also a Grade III marginal tissue recession with complete exposure of the buccal root and absence of attached gingiva in the first case. The key was correct diagnosis, timely treatment and keeping the goals realistic.

CONCLUSION

This case series emphasizes the need for careful evaluation of complicated cases of endo perio lesion with or without marginal tissue recession and exposure of bucal root where conventional therapy fails due to incorrect diagnosis. It also highlights the fact that functional restoration of the periodontium is one of the important aims of periodontal plastic surgery.

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Author Contributions
Sangeeta Singh – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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CASE SERIES OPEN ACCESS

Neglected ruptures of the patellar tendon: Repair options in a resource poor setting

Ikpeme A Ikpeme, Anthonia A Ikpeme, Innocent E Abang, Paul O Amah, Ahmed M Achaka

ABSTRACT

Introduction: Rupture of the patellar tendon is a disabling injury. Better results are reported in immediate repairs of fresh ruptures. In neglected ruptures, restoration of the structural and functional integrity of the extensor apparatus is difficult. This report highlights the use of an achilles osteotendinous autograft in the repair of an 8-month old neglected rupture of the patellar tendon; and the Mandelbaum technique with open tendon harvest in the repair of a 10-week old neglected rupture. Case Series: A 23-year-old male presented with inability to extend the left knee; eight months after a road traffic accident. Physical and radiological findings were consistent with a rupture of the left patellar tendon. The patient was offered tendon reconstruction using an Achilles osteotendinous autograft from the contralateral lower limb. One year later, he was pleased with the outcome. A 29-year-old army officer presented with pain, swelling and inability to extend his right knee since 10 weeks. Physical and X-ray findings were in keeping with a rupture of the right patellar tendon. The patient was offered patellar tendon reconstruction using the Mandelbaum technique. Twenty-eight weeks postoperatively, the patient had a range of motion of 0–105º in the right knee and has since returned to a functional professional and recreational lifestyle. Conclusion: Neglected ruptures of patella commonly yield suboptimal results following repair. In resource-poor settings, an autogenous achilles tendon technique as well as the Mandelbaum technique may prove beneficial in the treatment of this difficult condition.

Keywords: Patellar tendon, Neglected rupture, Achilles osteotendinous autograft, Mandelbaum procedure, Resource poor setting

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INTRODUCTION

Rupture of the patellar tendon is a disabling injury commonly seen in individuals under 40 years of age [1]. The injury is uncommon, can be easily missed in multi-injured patients, in the obese and in knees with hemarthrosis [1]. These ruptures usually occur at the inferior pole of the patella and better results are reported in immediate repairs of fresh ruptures [1, 2].

In our community, late presentation to orthodox care is common and the traditional bone setter is often the first to be consulted after injuries [3–5]. Neglected patellar tendon ruptures are thus only seen when the patient is unwilling to live with the disability that follows unorthodox treatment [4]. Neglected ruptures of

the patellar tendon (defined as ruptures presenting after six weeks) are often difficult to repair. Restoration of the structural and functional integrity of the extensor mechanism is difficult because of the associated muscle contracture, scar tissue formation, proximal patella migration and fibrous adhesions between the patellar and the femur [2, 6].

Many different techniques have been reported in the management of these conditions in long standing cases. End-to-end anastomosis of the tendon ends is often not feasible [2, 7]. Quadriceps lengthening, quadriceplasty, traction (preoperative and intraoperative), and external fixation with pins and wires are some techniques used to relocate the patella to its anatomic position. Autogenous graft augmentation using the fascia lata or hamstring tendons, allograft reconstruction using the Achilles tendon or patellar tendon are the commonly used reconstruction techniques for neglected ruptures. Prolonged postoperative immobilization, loss of knee flexion, residual weakness of the quadriceps and construct failure are complications associated with repair of a neglected rupture [7].

CASE SERIES

Case 1: A 23-year-old male presented to our out-patient department with complaints of inability to extend his left knee. He had been involved in a road traffic accident eight months earlier and had spent the interval in a traditional bone setter's practice.

Examination showed wasting of the left quadriceps muscles, a visible and palpable defect over the left knee, patella alta, loss of active extension and a passive range of motion of 10–60°. X-rays showed a proximally migrated patella with no fractures of the femur or tibia.

The patient was admitted for preoperative traction for two weeks and subsequently offered quadriceplasty and tendon reconstruction. Neither preoperative traction nor intraoperative elongation, and quadriceplasty were successful in restoring the patella to its exact anatomic location. Lack of tendon harvesters precluded the use of the hamstring tendons. The fascia lata was adjudged unhealthy following chronic inflammation from the traditional bone setters scarification. As we did not have access to allograft Achilles tendon graft, a decision to use autogenous Achilles tendon graft was made. Intraoperative findings were a proximally migrated patella, a 10-cm defect in the patellar tendon and marked fibrous adhesions involving the quadriceps femoris muscles.

A 1-cm central osteotendinous graft was harvested from the contralateral Achilles tendon. The defect at the donor site was closed with interrupted nonabsorbable sutures. The tendon graft was split into three slips, the central slip being thicker than the medial and lateral slips as described for the Achilles tendon allograft technique. A midline tunnel was drilled in the patella and the central slip passed through and anchored onto the quadriceps tendon from within outwards. The medial and lateral slips were anchored to either side of the patella and the medial and lateral tendinous expansions of the quadriceps tendon using non-absorbable interrupted sutures. The osseous pedicle was anchored into an appropriately sized window cut into the tibial tuberosity by two 3.5 mm cancellous screws. Quadriceplastly was performed. Postoperatively, patellar traction was continued for two weeks, the left lower limb was encased in a long leg cast after removal of traction and patient was discharged. He was seen at four weeks post discharge, the long leg cast was removed and an exercise programme commenced.

At 16th week postoperatively, the patient was pain free, had improved gait and an active range of motion of 5–95° in the left knee (Figures 1, 2). The ankle range of motion on the right side was normal and power of plantar flexion was grade 4+. He was subsequently lost to follow-up and re-appeared in the clinic one year later. The knee range of motion at this time was 3–105°. He could run and walk normally and was very pleased with his progress.

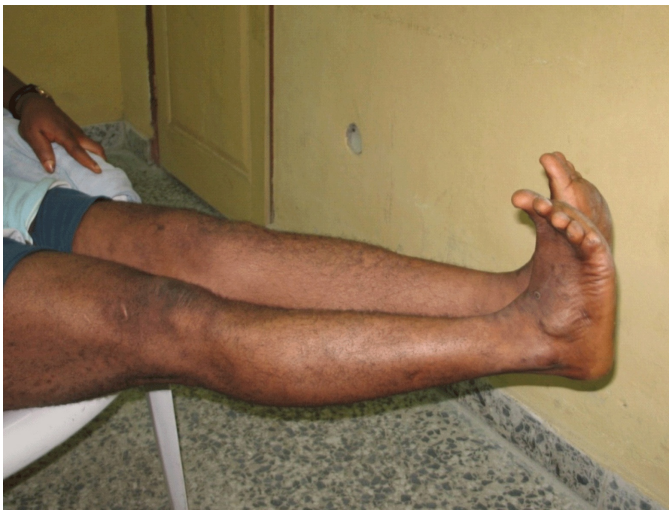

Figure 1: Showing range of active extension in both the knees. (case 1)

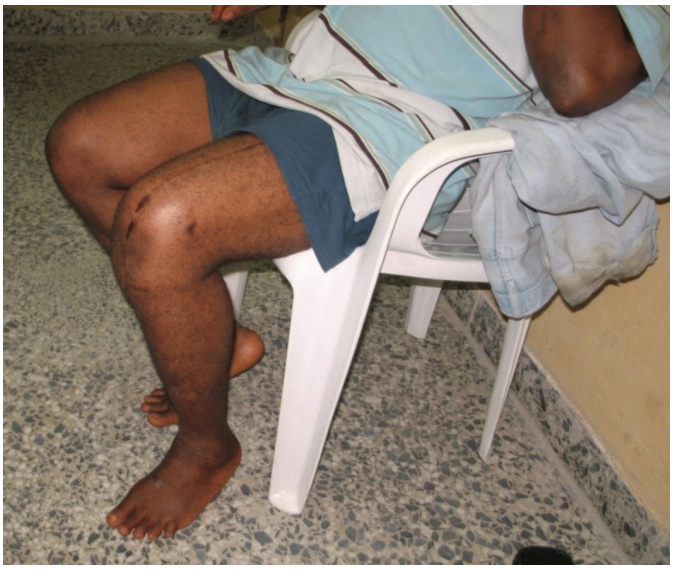

Figure 2: Range of active flextion in the knees. (case 1)

Case 2: A 29-year-old male army officer presented to the clinic with a 10-week history of pain, swelling and inability to extend the right knee following a fall onto the right knee while playing recreative basketball. Prior to presentation, he had sought unorthodox treatment with massage and hot fermentations and had lost the partial extension of the affected knee which was present in the early post injury period.

Examination revealed a swollen, mildly tender right knee, with a high riding patella and a distinct defect in the patellar tendon. The quadriceps femoris on the right side was wasted and patient had a range of active extension of 0° in the right knee. The range of right knee flexion was 110° and X-rays showed patella alta. An impression of neglected and mismanaged rupture of the right patella tendon was made.

The patient was admitted and offered patella tendon reconstruction using the Mandelbaum technique. The approach was an anterior midline approach. Intra-operative findings were a transverse rupture of the right patellar tendon and healthy semi-tendinosus and gracilis tendons. Operative technique involved minimal quadriceplasty, mobilization of the patella and relocation to its anatomic position. Owing to a lack of tendon harvesters, the skin flaps were raised until the semitendinosus and gracilis tendons were identified on the medial side. The tendons were traced to their musculotendinous junctions, sectioned at the junctions and their distal ends detached from the tibia. The tendons were then sutured end-to-end. Transverse holes were drilled across the patella and just below the tibial tubercle. The previously anastomosed semitendinosus and gracilis tendinous autografts were passed between the drill holes and used to reconstruct the extensor apparatus in a figure-of-eight configuration (Mandelbaum technique). A 0.5 guage malleable Kirschner wire was used to support the construct. The wound was closed in layers and a plaster of paris long leg cast was applied and worn for five weeks. The malleable Kirschner wire was removed six weeks post-operatively.

Postoperative management included immediate, non-weight bearing, bilateral, axillary crutch aided mobilization and graded range of motion physiotherapy exercises from the sixth week. The patient made a complete recovery with right knee range of motion being 0–105° at the last clinic visit 28th week after surgery. He has since returned to full military duties (including an international peace keeping tour of duty and a promotion) and has also returned to regular recreative basketball games.

DISCUSSION

Ruptures of the patella tendon usually follow long standing irritation. Chronic tendon degeneration due to repetitive micro trauma has been supported by histological evidence consistent with chronic inflammation and degeneration in ruptured tendon specimens [8]. Patella tendon rupture is usually unilateral when it results from traumatic athletic injuries, road traffic accidents and local steroid injections. When it occurs in association with systemic disorders such as systemic lupus erythematosus, chronic renal failure or diabetes mellitus, the rupture can be bilateral [9, 10]. The condition can also occur in posterior dislocations of the knee.

Suboptimal results have often been reported in the treatment of neglected injuries [1, 2, 6, 7]. In our setting, the combination of inappropriate unorthodox intervention and lack of basic requirements for tendon harvesting presented unique challenges. In our first patient, the fascia lata was also adjudged inappropriate for use due to evidence of chronic inflammation following scarifications by the traditional bone setter on the affected thigh. The combination of (possible) sub-clinical soft tissue infection (from scarifications and herbal fomentations to drain “evil-blood”) [3, 4] and poor nutrition can predispose to an unhealthy fascia lata. In our second patient, the challenges posed by inappropriate unorthodox interventions were obvious. In the immediate postinjury period and up to five weeks thereafter, the patient had weak extension of the right knee but lost that following massage and traditional ‘physiotherapy’. An apparently incomplete injury was converted to a complete rupture when the patient was presented to the clinic.

The treatment of neglected ruptures of the patellar tendon aims to relocate the patella in its anatomic position and recreate the Insall-Salvati ratio, ensuring mobility and correct tracking of the patella and restore the structural and functional integrity of the extensor apparatus [1, 2, 11]. This is difficult and the results are often sub-optimal [2, 6, 11]. Patellar relocation can be achieved by preoperative and intraoperative traction, quadriceplasty and a V-Y elongation of the quadriceps tendon. The use of external fixators and the Ilizarov technique have also been reported.

Tendon reconstruction involves the use of autogenous fascia lata or hamstring tendons; allograft patellar tendon or Achilles tendon in single or double graft techniques [2, 6, 7]. In our setting, there is no access to allograft tendons and autogenous hamstring or fascia lata grafts were not feasible in the first patient. An Achilles tendon autograft was thus the preferred option.

Autogenous Achilles tendon grafts can potentially present such complications as residual donor site rupture, infection and loss of plantar flexion. These may discourage the routine use of this technique. However, in resource-poor settings with no access to Achilles or patellar tendon allografts, no access to proper harvesting of an adequate length of hamstring tendons and where use of the fascia lata is precluded, our first case shows that an Achilles tendon autograft may be a feasible option. We believe the issues of donor site morbidity and infection can be addressed by ensuring harvest of only the central one cm of the tendon and meticulous attention to surgical technique. Graft take is improved by the inclusion of an osseous pedicle. In our first patient, the gait improved, range of motion in the left knee improved and plantar flexion power was grade

4+. We believe the outcome justifies our technique and may be useful in similar settings.

In our second patient who presented relatively earlier, the issues of sub-clinical infection did not exist. In non-resource challenged settings, the Mandelbaum technique would have been offered with the less risk of donor site morbidity using a tendon harvester. Its absence left the managing team with the option of an open tendon harvest. Though our results support this as a viable option when indicated in resource-challenged settings, the documented donor site morbidities such as infection must be borne in mind and appropriate steps taken to minimize the risk. Overall however, health systems and their managers must work towards the provision of an improved therapeutic armamentarium in our health sector. This will reduce the reported risks that attend orthodox intervention in the repair of this disabling injury. Health education to dissuade unorthodox intervention in orthopaedic maladies remains a credible intervention tool for improved outcomes.

To the authors' knowledge, an Achilles tendon autograft repair for neglected rupture of the patellar tendon has previously not been reported. We report our use of the technique of Achilles tendon autograft in the repair of an eight-month old neglected rupture of the patellar tendon in a 23-year-old male; and the use of the Mandelbaum procedure with open harvest of the hamstring tendons in the repair of a 10-week old neglected rupture of the patella tendon in a 29-year-old male. Both patients had spent eight months and 10 weeks, respectively seeking unorthodox care. In non-resource challenged settings a tendon harvester makes tendon harvest easier with less risk of donor site morbidity.

CONCLUSION

Neglected ruptures of the patellar tendon present unique treatment challenges especially in resource poor settings. An Achilles tendon autograft may prove beneficial in the treatment of this condition. The Mandelbaum technique using an open tendon harvest procedure is also a feasible option in these settings. Meticulous attention must be paid to the prevention of the possible complications that may occur following these techniques. Provision of appropriate intervention tools and health education to dissuade unorthodox interventions will help improve outcomes.

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Author Contributions

Ikpeme A Ikpeme – Substantial contributions to conception and design, acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Conflict of Interest

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CASE REPORT OPEN ACCESS

A patient with primary biliary cirrhosis and ulcerative colitis with progression to primary sclerosing cholangitis and colorectal cancer: A case report

Rui-Hua Shi, Bo Hao, Shun-Fu Xu, Qi-Yun Tang, Jian-Xia Jiang

ABSTRACT

Introduction: Ulcerative colitis (UC) is a chronic inflammatory disease of the colon characterized by intermittent exacerbations and remissions. It can be associated with primary biliary cirrhosis (PBC) or primary sclerosing cholangitis (PSC) and complicated with colorectal cancer (CRC). We describe a complicated case of associaton of PBC and chronic UC, eventually progressed to PSC and CRC. Case report: A 41-year-old female was diagnosed as PBC based on liver biopsy 10 years ago, and then UC was diagnosed too. Now symptoms of jaundice and abdominal pain aggravated. At last PSC and CRC were conformed. Conclusion: Prolonged Inflammatory bowel disease (IBD) accompanied with PSC in a patient has high risk of CRC. More exploration is needed to gain insight into relationship between IBD, PBD, PSC and CRC.

Keywords: Primary biliary cirrhosis, Ulcerative colitis, primary sclerosing cholangitis, colorectal cancer.

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INTRODUCTION

Primary billiary cirrhosis (PBC), primary sclerosing cholangitis (PSC), ulcerative colitis (UC) and colorectal cancer (CRC) are common diseases. PBC is marked by the slow progressive destruction of the small bile ducts (bile canaliculi) within the liver. When these ducts are damaged, bile builds up in the liver (cholestasis) and over time damages the tissue. This can lead to scarring, fibrosis and cirrhosis. PSC is a chronic liver disease caused by progressive immune-mediated inflammation and subsequent fibrosis of the bile ducts of the liver with the development of multiple strictures due to an intrinsic liver disease. The underlying cause of the inflammation is believed to be autoimmunity. Both PBC and PSC can be induced by autoimmune reactions. There are multiple similarities between these two diseases that may cause confusion or misdiagnosis, especially at the onset stage, including symptoms of fatigue, abdominal discomfort, pruritus and weight loss [1]. Diagnosis of PBC and PSC needs to be confirmed by liver biopsy.

UC is a chronic inflammatory disease of the colon characterized by intermittent exacerbations and remissions. It may be complicated with colon cancer or autoimmune-related extracolonic problems. It is not rare that PBC can be associated with UC and in such association occurrence of these diseases may not have particular order [2, 3]. Burnevich et al. reported that 24.1% of PBC patients has extrahepatic manifestations including UC [4]. It has also been reported that PBC can happen after proctocolectomy for ulcerative colitis [2].

There are many evidences that UC can be associated with PSC. Approximately, three quarters of patients with PSC have inflammatory bowel disease (IBD), e.g. UC or Crohn's disease [5] and 2–7.5% patients with UC have PSC [6–9]. It is, generally, accepted that approximately 5% of patients with UC will have the associated PSC.

We describe a complicated case of association of PBC and chronic UC, eventually progressing to PSC and CRC. Some special common mechanisms may be involved in the pathogenesis of these diseases.

CASE REPORT

A 41-year-old female was admitted with complaints of abdominal pain diarrhea and high fever for five days. The patient was diagnosed with PBC 10 years ago based on pathological examination of liver biopsy. Three years later, a colonoscopy examination proved a diagnosis of UC (Figure 1A). Symptoms including jaundice, pruritus and diarrhea persisted throughout the next seven years during which cholecystolithiasis developed in 2003 and relapsed in 2007. Four months before admission, the patient underwent hysterectomy and resection of right ovarian cyst along with right ovarian appendage in a local hospital. Rectovaginal fistula and rectal stenosis occurred one month after surgery. A metal stent was placed into the rectum. X-ray exhibited partial intestinal dilation with intermediate obstruction (beading). Magnetic resonance cholangiopancreatography (MRCP) showed typical appearance of PSC like diffuse strictures of both intrahepatic and extrahepatic bile ducts with dilation of the intervening areas and multiple bile duct stones in the common bile duct (Figure 1B–C). Severe abdominal pain and increasing skin icterus accompanied with high fever led us to take the patient for surgery. The laboratory test results of the patient are given in Table 1. A total colectomy and ileostomy were performed.

Peroperative findings: During surgery significant adhesions were observed in abdominal and pelvic cavity. Nodular cirrhosis of liver and splenomegaly was found. Biopsy from left external lobe of liver was taken for pathological examination. Ascending colon, cecum and small intestine were dilated with thickening and swelling of bowel wall. A hard mass of 3x3x3 cm invading the submucosa was found in ascending colon near the hepatic flexure which was causing intestinal obstruction. Distal colon and upper part of rectum were thickened and stiff.

Pathological results and final diagnosis: Histopathological examination of colon showed mucosal atrophy with loss of crypts and distortion of the mucosal architecture. Some of the crypts were shortened and branching. Transmucosal inflammation with basal plasmacytosis, lymphocytosis and eosinophilia were evidenced by the presence of neutrophils infiltrating the walls of some crypts. Thickening of mucosa muscularis layer and hyperplasia of fibrotic and fatty tissue in the submucosal layer indicated late stage of ulcerative colitis (Figure 2A). Grade II colonic adenocarcinoma was found which was invading all the layers of colon from epithelium to serosa to the surrounding connective tissue (Figure 2B). Lymph nodes metastasis was found positive in one out of 10 lymph nodes. The final diagnosis was UC concomitant with PSC complicated by colonic adenocarcinoma, liver cirrhosis, hypersplenism, post-cholecystectomy, post-hysterectomy, and post-unilateral adnexectomy. The patient was regularly followed-up and was given Pentasa, 1 g, qid, po; UDCA 250 mg, bid po. and UDCA without discomfort.

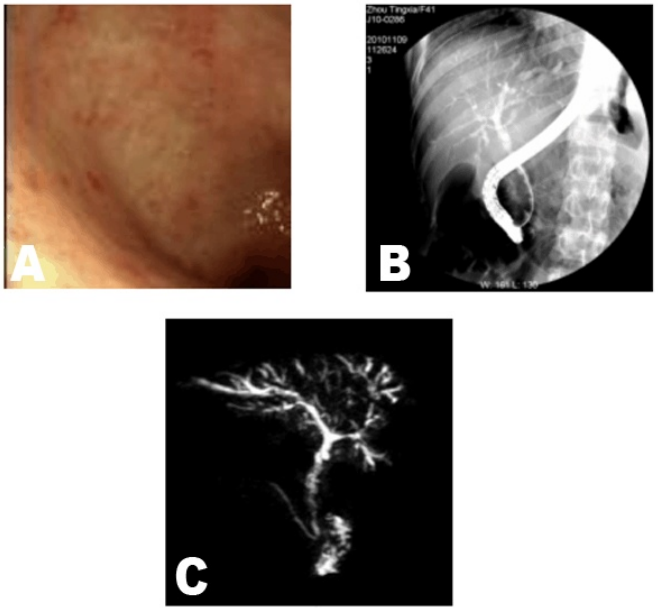

Figure 1: (A) Colonoscopy picture used for diagnosis of UC in 2004, (B) image of MRCP that showed multiple bile duct stone in the lower part of common bile duct, (C) Retrograde cholangiopancreatography showed that both intra- and extra-hepatic bile duct, exhibited segmental and diffused stenosis and expansion, which conforms with the diagnosis of PSC. Stones were shown in common bile duct.

DISCUSSION

We reported a complicated, multi-disease case with long-duration of clinical manifestations. Although certain diseases discussed here may be an isolated event,

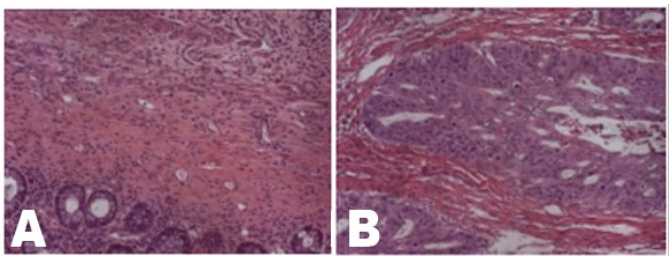

Figure 2: Pathologic sections for UC and CRC diagnosis. (A) Section from colon tissue showed late stage of UC, (H&E, x200) (B) Section of colon neoplasms that invaded whole layer of submucosa (H&E, x200).

Table 1: Laboratory test reports of the patient.

Lab examination	Results
Blood routine test	WBC 2.6x10^3/μL, NE 66.00%, RBC 2.70x10^6/μL, Hb 7.7 g/dL, PLT 99x10^3/μL
Urine routine test	Bilirubin 2+
Electrolyte	Ca^{2+} 2.13 mmol/L, P^{+} 0.38 mmol/L, Mg^{2+} 0.47 mmol/L
Biochemical markers	Albumin 2.71 g/dL, SGPT 80.4 U/L, SGOT 115.8 U/L, GGT 167.6 U/L, ALP 1205.8 U/L, Bilirubin (total) 341.0 mol/L, Bilirubin (indirect) 185.10 mol/L, Alb/Globulin 0.8
Coagulating function	PT 17.9 s, APTT 59.6 s, Fibrinogen 60 mg/dL, PT-INR 1.65, TT 25.5 s
ESR and CRP	ESR 7 mm/1st h, CRP 60 mg/L
Immunoglobulin and complement anti-ENA antibodies	IgG 295 mg/dL (7.0–16 mg/dL), C3 0.5x10^3 μg/mL (0.9–1.8x10^3 μg/mL), anti-ENA antibodies was negative
Tumor markers	CEA 19.54 ng/mL (0–4.3 ng/mL), CA19.9 76.33 U/mL (0–39 U/mL)

most of the diagnoses reported in literature have shown a pattern of more or less interconnection with a common mechanism for disease progression.

Liver biopsy clearly indicated that the onset of all symptoms in this case started from PBC and progressed to UC and further to PSC. It is unclear whether PBC can directly progress to PSC. Not many cases of PBC progression to PSC have been reported. Jeevagan et al. reported one case recently in which PBC and PSC were diagnosed in the same patient but failed to clarify whether it is just an overlapping phenomenon or progression of one disease to another [10]. In a large study in Sweden with 1500 UC patients, it was found that 5% patients had increased serum alkaline phosphatase and 3.7% had evidence of PSC [11]. The same study showed that 95% patients who had PSC were found to have UC. In another study, 48/336 (48%) patients with UC had evidence of hepatobiliary pathology and 4% patients with UC had PSC [12]. In our case, the patient received ERCP twice for removing stones in the bile duct. It is reported that cholecystolithiasis was found in 23/41 (56%) UC patients, of which 7/23 (30%) were missed on cholangiography and detected only by cholangioscopy [13]. Major investigations for stone detection include CT scan, sonography and cholangiogram. Endoscopic therapy can provide drainage of bile ducts, removal of stones and/or temporary relief from obstruction [14].

After the prolonged duration of UC, the possibility of colorectal carcinoma should be highly suspected. The patient was found to have CRC during the surgical operation, which had already invaded through the whole layer including the serosa and showed lymph node metastasis.

UC-PSC shows unique colonoscopy features and are associated with more frequent colorectal neoplasm development and poor prognosis [15]. The overlap of UC and PSC constitutes a higher risk of developing colorectal dysplasia/carcinoma than UC patients without PSC. Prolonged disease duration of IBD is another important risk factor for CRC [16]. Eaden's meta-analysis has shown that the risk for CRC in UC patients is 2% at 10 years, 8% at 20 years and 18% at 30 years of disease duration [17]. The onset of CRC was significantly younger in patients with IBD and PSC [7]. Terg R et al. analyzed the prevalence of PSC in 1,333 patients with UC and the risk for developing colon cancer. Seven out of 39 (18%) patients with PSC developed colorectal carcinoma compared with 2/78 (2.6%) in the control group (p=0.006). The cumulative risk of colorectal carcinoma was 11% and 18% after 10 and 20 years in the PSC group compared with 2% and 7% in the control group, respectively (p=0.002) [9]. It is highly recommended to have early and/or regular colonoscopy screening for IBD patients with or without PSC.

CONCLUSION

Prolonged inflammatory bowel disease, accompanied with PSC is not an uncommon disease that has high risk of CRC. Close follow-up of patient with regular colonoscopy for neoplasm detection will help in the early diagnosis and extension of life span. Additional research studies such as prospective studies are needed to gain insight into disease evolution and relationship between IBD, PBC, PSC and CRC. Additionally, exploring when CRC screening should begin in IBD and PSC patients would be a relevant topic.

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Author Contributions

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Bo Hao – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Shun-Fu Xu – Substantial contributions to conception and design, Acquisition of data, Revising the article critically for important intellectual content, Final approval of the version to be published
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Jian-Xia Jiang – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

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CASE REPORT OPEN ACCESS

Sarcomatoid bladder carcinoma: A rare metastatic disease of small bowel

Tafadzwa Makarawo, Joshua Phillips, Jonathon Eaton, George Kondratowicz

ABSTRACT

Introduction: Small bowel obstruction secondary to metastatic disease is a rare presentation with most current evidence being limited to case reports. Previous evidence described breast, lung and melanoma as the most common underlying primaries presenting with this entity. Bladder carcinoma rarely metastasizes to the gastrointestinal tract, and if present, does not typically cause small bowel obstruction. Case Report: A 62-year-old female with a history of sarcomatoid bladder carcinoma that had been managed initially by transurethral endoscopic resections and subsequently by a radical cystectomy presented a year later with small bowel obstruction. Extensive work-up for the source of the obstruction was inconclusive and after failed conservative management, the patient underwent an exploratory laparotomy. At laparotomy, she was found to have a small bowel intussusception secondary to a tumor. The small bowel resection specimen was later identified as a sarcomatoid carcinoma, resembling the previously resected bladder tumor. Conclusion: We present a rare case of a patient presenting with small bowel obstruction secondary to metastatic disease from a variant of bladder cancer—the sarcomatoid bladder carcinoma.

Keywords: Metastatic bladder carcinoma, Bowel obstruction

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INTRODUCTION

Metastatic disease to the intestine is common although the incidence can vary significantly depending on the primary tumor type [1]. However, small bowel metastasis rarely present with obstruction, with only 56 cases being reported up to 2012. The most common primary sites include breast, lung, renal and ovarian cancer [1]. Even less common is the presentation of intestinal obstruction secondary to metastasis from a bladder cancer primary, which has only been reported once in English literature, the previous case being a large bowel obstruction [2]. Here we present a rare case of small bowel obstruction secondary to a small bowel metastasis from a bladder primary.

CASE REPORT

A 62-year-old Caucasian female was admitted with colicky abdominal pain and vomiting. The patient's previous history was significant for sarcomatoid

transitional cell carcinoma of the bladder for which she had undergone multiple local endoscopic resections but subsequently had a radical cystectomy with ileal conduit formation for muscle invasive (pT2) disease. Following surgery, the final cystectomy specimen showed no residual tumor. She did not receive adjuvant chemoradiotherapy.

Upon admission, her abdominal pain was initially managed conservatively. Her symptoms subsequently resolved within 48 hours and she was discharged home. However, she again presented four days later with further abdominal pain and vomiting. An abdominal computed tomography (CT) scan demonstrated mildly dilated small bowel loops but no obstructing lesion. Her symptoms were attributed to adhesions and once again managed conservatively with subsequent discharge home.

Approximately three weeks later, she presented again for a third time with obstructive bowel symptoms. X-ray showed a normal gas pattern although on this occasion her inflammatory markers were mildly elevated. Upper GI endoscopy only revealed a non-occlusive benign duodenal polyp while contrast small bowel studies and an oral and rectal contrast-enhanced CT scan demonstrated prominent small bowel loops without an obstructing lesion.

With her symptoms becoming protracted, a decision was made to proceed with laparotomy. At the time of surgery, the small bowel obstruction was found to be due to a small bowel intussuception with a jejunal tumor as the lead point. A small bowel resection was therefore performed with a primary end-to-end anastomosis. The patient made a steady postoperative recovery and was eventually discharged home one week later.

Histology of the resected jejunal tumor revealed a six-cm submucosal nodule with spindle cell areas and occassional multinucleated cells. Immunohistochemistry of the tumor showed presence of vimentin, a feature of sarcomatoid malignancy. These findings correlated with the resected bladder carcinoma, confirming it as a metastatic tumor.

On most recent follow-up (one month after surgery), the patient was found to have extensive brain and liver metastases deeming her with a poor prognosis.

DISCUSSION

Bladder cancer is the fifth most common cancer worldwide with a reported incidence of 104,400 in the European Union in 2006 and 68,810 cases documented in the United States in 2008. In the industrialized countries, more than 90% bladder cancers are transitional cell carcinomas, while in the the developing countries, 75% are squamous cell carcinomas. Sarcomatoid bladder carcinoma is a rare form of bladder cancer, with an incidence of 0.31% [3].

Sarcomatoid carcinomas are biphasic tumors that by definition contain an epithelial component adjacent to a mesenchymal component. The first detailed report of sarcomatoid carcinoma of the urinary bladder was described in an article by Robson et al. in 1935 [4]. Histologically, sarcomatoid carcinomas contain spindle-shaped cells with abundant eosinophilic cytoplasm and atypical, bizarre, hyperchromatic nuclei within the stroma (Figure 1) [5]. Although the behaviour of sarcomatoid carcinomas is still under scrutiny due to their rarity, it is well known that they usually present with a high degree of malignancy [5], and have presented with metastatic disease even in non-muscle invasive (T1) primaries [3].

Bladder carcinomas, as with most tumors of the genitourinary system most frequently spreads to lymph nodes (59%), liver (47%), lungs (45%) and bones (32%).

Reported cases of bladder metastases to the small bowels are rare [6–8], with none described in current English literature. Interestingly, the converse, i.e. metastatic spread of small bowel tumors to the bladder, causing urological symptoms is equally uncommon with only one reported case in current literature [9].

Small bowel obstruction is a rare presentation of metastatic disease, with most current evidence limited to case reports [1]. Published evidence suggests that breast cancer is by far the most common distant primary site causing with small bowel obstruction, secondary to isolated metastases to the bowel wall (47% cases) [1]. The next most common primaries causing small bowel obstruction are lungs (8%) and malignant melanoma (11%). Most case reports have described the mechanism of bowel obstruction in these patients as being related to intussusception, with the metastatic tumor being the lead point. To our knowledge, this is the first case reported in English literature of bladder metastasis to the small bowel causing intussusception and bowel obstruction. The only other simular case caused large bowel obstruction [2].

The diagnosis of small bowel obstruction due to metastatic disease poses a significant challenge. As our case demonstrated, the rarity of the condition caused by bladder metastases lead to a low index of suspicion, plus the presenting symptoms can often be non-specific in nature and as a result the diagnosis may be significantly delayed.

Our patient developed metastatic disease, despite seemingly successful aggressive treatment of her bladder cancer with no evidence of residual tumor in the final bladder specimen on histology. This once again

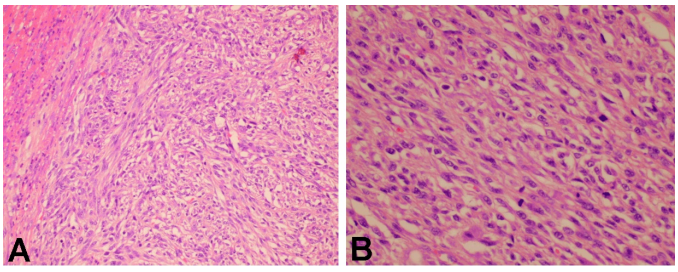

Figure 1: Histological sections of (A) resected bladder carcinoma (H&E, x200), (B) and resected small bowel tumor (H&E, x200). Similarities in features between the two sections include spindle celled areas and occasional multinucleated cells, consistent with sarcomatoid carcinoma.

highlights the aggressive nature of sarcomatoid carcinomas. The presence of this rare but aggressive form of bladder cancer as a primary should, therefore, raise the index of suspicion of subsequent metastatic sequelae, particularly in cases with a clinical picture suggestive of bowel obstruction.

CONCLUSION

Bowel obstruction due to metastasis from bladder cancer is an extremely rare entity. In the presence of a history of sarcomatoid bladder primary, the possibility of small bowel metastatic disease should be considered in patients who present with bowel obstruction.

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Author Contributions

Tafadzwa Makarawo – Substantial contributions to conception and design, acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Joshua Phillips – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published
Jonathon Eaton – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
George Kondratowicz – Substantial contributions to conception and design, Acquisition of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

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Conflict of Interest

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CASE REPORT OPEN ACCESS

Choledochal cyst with portal hypertension: A case report

Rahul Roy, Jyoti Bansal, Yeshwanth Rajagopal,
Rajendra Mandia, Raj Kamal Jenaw

ABSTRACT

Introduction: Choledochal cysts in adults are commonly associated with hepatobiliary pathology and complications of previous cyst related procedures. Portal hypertension is a rare complication of choledochal cyst. The treatment of choledochal cyst complicated by portal hypertension has evolved from internal drainage of cysts to single stage excision of cyst with bilio-enteric anastomosis. Case Report: A 15-year-old female presented with typical triad of abdominal pain, abdominal lump and jaundice. Magnetic resonance cholangiopancreatography (MRCP) was suggestive of type-I choledochal cyst with portal hypertension. An upper gastrointestinal endoscopy revealed grade 1 esophageal varices with proximal gastropathy. Intraoperatively, the posterior wall of the choledochal cyst was densely adherent to the portal vein and hence a partial excision of cyst with stripping of the mucosa of the posterior wall of the cyst along with Roux-en-Y hepaticojejunostomy was done. Conclusion: Single stage excision of choledochal cyst with bilio-enteric anastomosis is the treatment of choice of choledochal cyst with portal hypertension. Portal decompression is reserved for cases with extensive collaterals in the hepatoduodenal ligament.

Keywords: Complicated choledochal cyst, Portal hypertension

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INTRODUCTION

Choledochal cyst is predominantly a disease of childhood. However, about 20% cases are diagnosed in adults. With the recent advances in imaging technology the incidence of choledochal cyst is increasing. Choledochal cyst in adults are commonly associated with hepatobiliary pathology and complications of previous cyst related procedures.

Portal hypertension is a rare complication of choledochal cyst. The treatment of choledochal cyst complicated by portal hypertension has evolved from internal drainage of cyst to single stage excision of cyst with bilioenteric anastomosis. Portal decompression is reserved for cases with extensive collaterals in the hepatoduodenal ligament [1, 2].

Here we report a case of choledochal cyst with portal hypertension.

CASE REPORT

A 15-year-old female was presented with typical traid of abdominal pain and abdominal lump for one year and

jaundice for eight months. For last two months, she also had pruritus with passage of clay colored stools. There was no previous history of acute cholecystitis, pancreatitis, hematemesis or melena. On biochemical investigations, hemoglobin was 8.3 g/dL, serum bilirubin 6.3 mg/dL, serum SGOT 258 U/L, serum SGPT 105 U/L, serum alkaline phosphatase 711 IU/L and albumin-globulin ratio was 1:1.2. Viral markers for hepatitis B and hepatitis C were negative. Abdominal ultrasonography showed a large cystic lesion of size 11.5x13 cm in epigastric region, hepatosplenomegaly with heterogenous coarse echotexture of liver, dilated intrahepatic biliary radicle (IHBR) and common bile duct (CBD). Proximal CBD measured 2.2 cm with non-visualization of distal portion.

On further investigating the patient, magnetic resonance cholangiopancreatography (MRCP) showed a large cystic dilatation of CBD measutring 11.8x11.6x11.6 cm (type-I choledochal cyst) with minimum sludge in dependent position likely to be choledochal cyst. Also seen were dilated IHBR and hepatic ducts, nodular liver, splenomegaly, displaced portal vein with recanalization of left umbilical vein and ascites (Figure 1). An upper gastrointestinal endoscopy revealed grade 1 esophageal varices with proximal gastropathy. Serum-ascites albumin gradient (SAAG) was >1.1 g/dL wich was suggestive of portal hypertension.

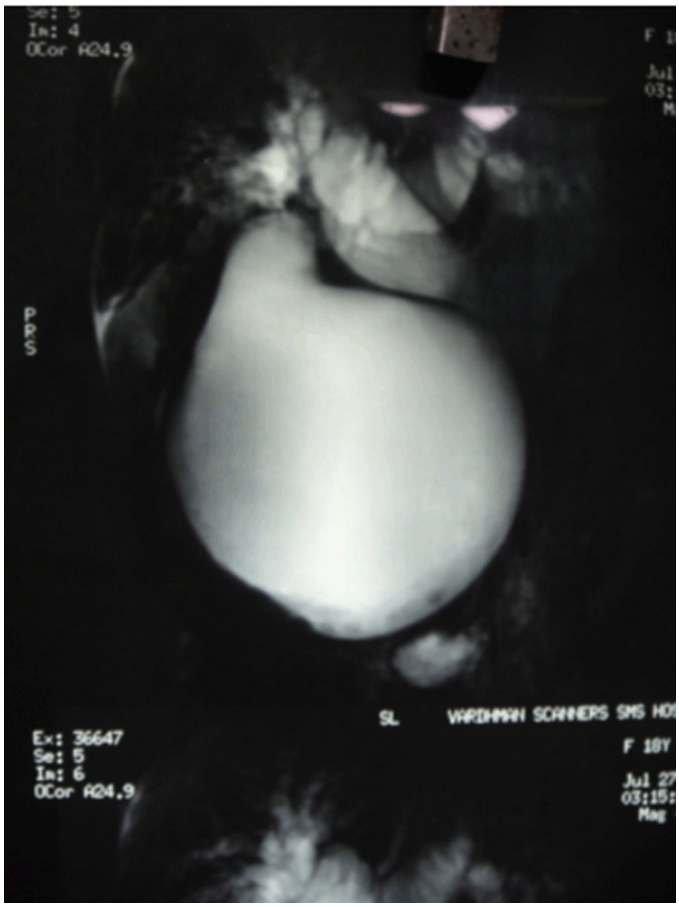

Figure 1: Magnetic resonance cholangiopancreatography (MRCP) suggestive of choledochal cyst and dilated intra-hepatic biliary radicle.

A preoperative diagnosis of choledochal cyst with portal hypertension was made and single stage operative procedure which included excision of the choledochal cyst with bilioenteric anastomosis was planned. Intraoperatively, large focal segmental dilation of CBD below the cystic duct (type-IB choledochal cyst) displacing the portal vein on left with dense adhesions between the portal vein and posterior wall of the choledochal cyst, and splenomegaly with multiple collaterals in the hepatoduodenal ligament were evident. The posterior wall of the choledochal cyst could not be separated from the portal vein; so partial excision of the cyst with stripping of the mucosa of the posterior wall of the cyst along with a Roux-en-Y hepaticojejunostomy was performed. The postoperative period was uneventful and the histopathological examination was suggestive of an inflamed choledochal cyst. At 16 months of follow-up the patient was well with complete regression of esophageal varices.

DISCUSSION

With advancement of imaging modality, incidence of adult choledochal cyst is on rise. Incidence in Asia is somewhat higher than in western countries. The reason for this geographical difference is still unclear [1, 2]. There is also an unexplained female preponderance with female:male ratio commonly reported as 4:1. The most widely accepted hypothesis regarding etiology is an anomalous arrangement of the pancreaticobiliary ductal junction [3, 4]. Choledochal cyst is a disease of infancy and childhood but about 20% are not diagnosed until adulthood [5–7]. Choledochal cysts in adults are more commonly associated with hepatobiliary pathology and complications of previous cyst related procedures [1, 6, 7]. The complications include cystolithiasis, hepaticolithiasis, cholangitis, calculous cholecystitis, pancreatitis, pancreatic duct abnormalities, malignancy and portal hypertension. Cystolithiasis is the most frequent complication in adults with choledochal cyst with a prevalence rate ranging from 2–72% [7]. The treatment of choice of choledochal cyst is excision of cyst with bilioenteric anastomosis. In conditions where complete excision of cyst is not possible due to adhesion with vital structures, partial excision of cyst with stripping of mucosa of the part of cyst left *in-situ* can be done as stripping of the mucosa removes the tissue with malignant potential.

Portal hypertension is a rare complication of long standing choledochal cyst manifested clinically as hepatosplenomegaly, jaundice, hematemesis, melena and ascites. Portal hypertension in patients of choledochal cyst may be due to extrahepatic biliary obstruction leading to secondary biliary cirrhosis, recurrent inflammation leading to portal vein thrombosis, direct compression of portal vein by choledochal cyst or associate congenital hepatic fibrosis in patients with Caroli disease [4, 7–9].

Choledochal cyst complicated by portal hypertension should be differentiated from portal biliopathy. Portal

biliopathy, a recent terminology, has been used to describe changes in the bile duct due to cavernous transformation in patients with portal hypertension. Such changes are more common in patients with extrahepatic portal vein occlusion. These biliary abnormalities are classified as varicoid, fibrotic or mixed. In the varicoid type there is irregular contour of the bile duct as a result of multiple smooth extrinsic compression of the cavernoma clearly seen in MRCP or magnetic resonance angiography. In the fibrotic type magnetic resonance scans show localized strictures with proximal dilatation [10].

The various case reports and case series previously documented in literature are summarized in Table 1.

Table 1: Summary of documented cases of choledochal cysts with portal hypertension

Author	Age	Sex	Duration of symptom	Symptom	Operation	Result	Follow up	Cause of death
Gillis et al. [8]	2 yr	F	NA	Recurrent GI bleeding	Choledochocysto-jejunostomy	R	3 yr	
Gillis et al. [8]	5 yr	F	NA	Hepatosple-nomegaly, massive GI bleed	Choledochocysto-jejunostomy	R	10 mths	
Fonkalsrud et al. [8]	5.5 yr	F	NA	Massive GI bleed	Splenorenal shunt and choledochocystojej-unostomy	R	1 yr	
Duckett et al. [8]	6 yr	F	NA	Massive GI bleed, ascites	None	D		NA
Martin et al. [8]	8 mths	F	9 mths	Massive GI bleed	None	D		NA
Martin et al. [8]	2 yr	F	NA	NA	Choledochocystojej-unostomy	R	3 yr	
Martin et al. [8]	14 yr	F	2 yr	Massive hematemesis	Choledochocystojej-unostomy	R	4 yr	
Rao et al. [4]	11 yr	F	6 mths	Jaundice, lower GI bleed, lump abdomen	Cyst excision , jejunal loop inter-position hepatico-duodenostomy	R	3 mths	
Rao et al. [4]	12yr	M	6 mths	Jaundice, hematemesis	Roux-en-Y cystojejunostomy	R	1 yr	
Rao et al. [4]	11 yr	M	9 yr	Jaundice, hematemesis, hepatosple-nomegaly	Splenectomy with a vascular shunt	D		Fatal hemate-mesis
Rao et al. [4]	11 yr	M	3 mths	Jaundice, hepatosple-nomegaly	Cyst excision and Roux-en-Y hepaticojejunosto-my	R	6 mths	
Singh et al. [7]	50 yr	F	1 yr	Jaundice, hepatosple-nomegaly	Cyst excision and Roux-en-Y hepaticojejunosto-my	R	NA	
Saluja et al. [9]	40 yr	F	2 yr	Jaundice, hepatosple-nomegaly , ascites	Cyst excision and Roux-en-Y hepaticojejunosto-my	R	1 yr	
Saluja et al. [4]	66 yr	M	7 yr	Jaundice, ascites, melena	None	D		Liver failure
Saluja et al. [4]	49 yr	M	9 mths	Jaundice, hepatomegaly	None	D		Septicemia

Abbreviations : yr - years, mths - months, M - male, F - female, R - recovered, D - death, GI - gastointestinal, NA - data not available

Gillis et al. reported two cases of choledochal cyst with portal hypertension in which choledochojejunostomy was performed with regression of features of portal hypertension at 10 months follow-up in one case and three years in the second case [8]. Martin et al. reported three cases of choledochal cyst with portal hypertension managed by them in which choledochojejunostomy was done in two cases with regression of symptoms of portal hypertension at three years follow-up in one case and four years in the other. In the third case, surgery was refused by the parents and the patient died [8].

Fonkalsrud et al. reported a case in which a choledochal cyst was missed on initial evaluation and a splenorenal shunt was done. Subsequently, the expected fall in portal pressure did not occur and on further exploration of abdomen a choledochal cyst was found and a choledochojejunostomy was performed with an immediate fall in portal pressure. At one year follow-up there was a complete regression of esophageal varices [8]. The case emphasized that a shunt procedure for portal decompression in complicated choledochal cyst with portal hypertension will not lead to regression of portal hypertension and only excision of cyst will cure the portal hypertension.

Rao et al. presented a review of four cases of choledochal cyst with portal hypertension managed by them. In the first case, cyst excision with isolated jejunal loop interposition hepaticoduodenostomy was done with gradual regression of esophageal varices and congestive gastropathy at three months follow-up. In the second case, Roux-en-Y cystojejunostomy was done with regression of esophageal varices at three months follow-up. In the third case, there were extensive collaterals around the porta and splenectomy with a vascular shunt between inferior mesenteric vein and renal vein was done but the patient died while waiting for a definitive surgery due to fatal episode of hemetemesis. In the fourth case, Roux-en-Y hepaticojejunostomy was done with complete resolution of varices at six months follow-up [4].

Saluja et al. repoted three cases of choledochal cyst with portal hypertension managed by them. In the first case, Roux-en-Y hepaticojejunostomy was done and patient was well at one year follow-up. The second case was associated with alcoholic liver disease with Child Class C cirrhosis and the patient died of liver failure. In the third case, the patient initially underwent an ERCP with stenting followed by a repeat ERCP with removal of multiple stones after lithotripsy. Three months later patient developed cholangitis with renal failure and the patient died nine months after the diagnosis [9]. Singh et al. reported a case of choledochal cyst with portal hypertension managed by them in which Roux-en-Y hepaticojejunostomy was done [7].

In a retrospective study of 144 patients with choledochal cysts managed between January 1989 and June 2004 at a tertiary level referral hospital in North India, six patients had portal hypertension. Cyst excision was performed successfully in three out of six patients. In two patients an internal drainage was resorted to because of excessive bleeding from the collaterals in the hepatoduodenal ligament. One patient did not report for definitive surgery after a percutaneous biliary drainage for recurrent severe cholangitis [11].

It is evident from the review of literature that treatment of choledochal cysts complicated by portal hypertension has evolved from internal drainage of cysts to single stage excision of cyst with bilioenteric anastomosis. Endoscopic drainage may be considered as a temporary measure in patients who are unfit for surgery. In the presence of hypervascularity of the hepatoduodenal ligament and pericholedochal varices an attempt for cyst excision should be made rather than a shunt procedure for portal decompression as a shunt will not cure the portal hypertension and a second stage surgery for the excision of choledochal cyst will still be required. However, in the presence of extensive collaterals in the hepatoduodenal ligament, which is more commonly seen in associated portal vein thrombosis, portal decompression in the form of porto-systemic shunt should be done first followed by cyst excision 6–12 weeks later [6, 7, 11]. It should be kept in mind that in patients with Child Class C status a shunt may deteriorate liver function by diverting the portal blood flow and hence liver transplantation should be offered to such patients.

CONCLUSION

Treatment of complicated choledochal cyst has evolved over the years from internal drainage to single stage excision. Single-stage excision of cyst with bilioenteric anastomosis is the treatment of choice for choledochal cyst with portal hypertension. In cases, where complete excision of cyst is not possible, partial excision of cyst with stripping of mucosa can be done regression of varices without any evidence of recurrence is a pointer for preference towards single stage procedure.

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Rahul Roy – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Jyoti Bansal – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Yeshwanth Rajagopal – Conception and design, Acquisition of data, Analysis and interpretation of data,

Drafting the article, Critical revision of the article, Final approval of the version to be published
Rajendra Mandia – Acquisition of data, Critical revision of the article, Final approval of the version to be published
Raj Kamal Jenaw – Acquisition of data, Critical revision of the article, Final approval of the version to be published

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CASE REPORT OPEN ACCESS

Transmesosigmoid hernia with small bowel strangulation

Ji Won Kim, Dae Hyun Yang

ABSTRACT

Introduction: Internal herniation of the small bowel is difficult to diagnose before surgery. Delayed laparotomy is related to high morbidity and mortality. We present a rare case of a transmesosigmoid hernia to demonstrate the importance of high suspicion and early laparotomy. Case Report: A 44-year-old female patient had severe abdominal pain of acute onset. A CT scan at six hours after onset of the symptom showed a dilated small bowel loop of diminished attenuation. Her severe pain was not controlled even with narcotics. An emergency laparotomy eight hours after onset revealed about a 150 cm strangulated closed loop of the mid small intestine, herniated through a small mesenteric defect of the sigmoid colon. The defect in the mesosigmoid was about three cm in diameter without a sac, and functioned similarly to a napkin ring; therefore the reduction of the herniated closed intestine loop was difficult without division of the intestine. The patient recovered well after resection anastomosis and closure of the mesenteric defect. Conclusion: Congenital internal hernia is rare, but early development of incarceration and strangulation is associated with increased morbidity and mortality. Early suspicion and urgent or emergent laparotomy or laparoscopic exploration and surgical management are necessary.

Keywords: Transmesenteric hernia, Sigmoid colon, Internal hernia, Transmesosigmoid hernia

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INTRODUCTION

Small intestinal obstruction is a frequent cause of surgical emergency. The most common cause of small intestinal obstruction is postoperative adhesion. Internal hernia is a rare cause of intestinal obstruction with nonspecific symptoms and signs, and high suspicion is therefore necessary in the evaluation of abdominal symptoms suggestive of intestinal obstruction. Although abdominal computed tomography (CT) scan is believed to facilitate the diagnosis and early surgical exploration, the diagnosis of internal hernia remains a challenge for surgeons and radiologists. We present a case of internal hernia, through a congenital defect of the sigmoid mesocolon in a 44-year-old female who presented at our hospital with acute cramping abdominal pain.

CASE REPORT

A 44-year-old female patient visited the emergency department of our hospital with a 2-hour history of severe cramping abdominal pain. It began as low

abdominal pain after dinner and developed as whole abdominal pain. Also, she was experiencing nausea and the desire to defecate, and vomited once after a small amount of ingested food. She had a history of laparotomy for tubal ligation 20 years prior. Her vital signs were stable with a blood pressure of 120/50 mmHg, body temperature of 36.1°C, pulse rate of 66/min, and respiratory rate of 14/min. She was acutely ill-looking and complained of intractable abdominal pain. Abdominal physical examination revealed generalized tenderness. There was no pronounced rebound tenderness or guarding. A plain abdominal X-ray showed a mild ileus without significant bowel dilation (Figure 1). A hematologic profile at the time of hospitalization showed only an increased white blood cell count of $12.94x10^3/mm^3$ with neutrophil predominance of 78.4%, and the results of blood gas analysis and serum bilirubin, creatinin and amylase were within normal limits. Abdominal CT scan demonstrated ascites, dilated small bowel loop with a cut-off at the level of mid-ileum, and intestinal wall thickening (Figure 2). The cause of the abdominal pain was difficult to identify. The abdominal pain was not controlled even with narcotics and it was becoming worse. The second complete blood count test at four hours interval showed a more increased white blood cell count of $2.05x10^3/mm^3$ with neutrophil predominance of 94.2%. With the suspicion of bowel strangulation, an emergent laparoscopy was performed. Intraperitoneal evaluation was performed using a laparoscope inserted through the lower abdominal region. Hemorrhagic ascites and a dark strangulated dilated small intestinal loop were found; we, therefore, converted to open laparotomy of the lower midline incision. A long loop of small bowel with strangulation had herniated through a small defect of the sigmoid colon mesentery from left to right (Figure 3). There was no hernia sac. The strangulated small bowel loop of about 150 cm, of which the distal portion was 170 cm from the ileocecal valve, was entrapped at the hernia defect, mimicking the shape of a napkin ring. The defect was about 3x3 cm and was located near the base of the mesentery of the sigmoid colon. The incarceration could not be relieved without resection of the herniated bowel. After anastomosis of the bowel, the mesocolic defect was repaired by primary closure with nonabsorbable sutures. The patient had an uneventful postoperative course and was discharged on postoperative day 7.

DISCUSSION

Internal hernia is defined as a protrusion of an organ, usually the small intestine, through a normal or abnormal aperture within the abdominal cavity. The incidence of internal hernia is estimated to account for approximately 1–6% of intestinal obstructions [1]. Sigmoid mesocolon hernia is an uncommon type and estimated to account for approximately 6% of internal hernia [2, 3]. Benson classified sigmoid hernia into three types [4].

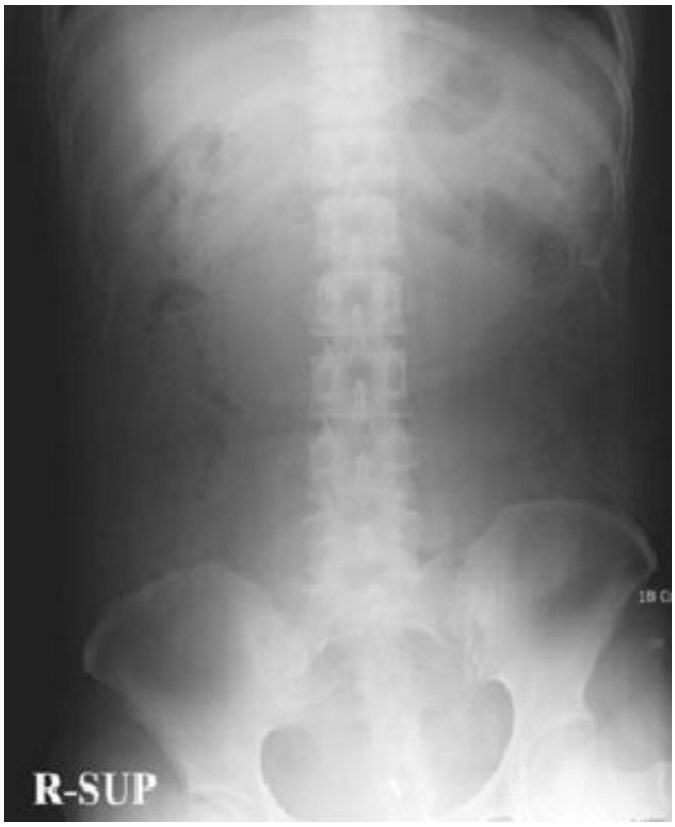

Figure 1: Plain abdominal X-ray film showed mild ileus without prominent dilation of small bowel.

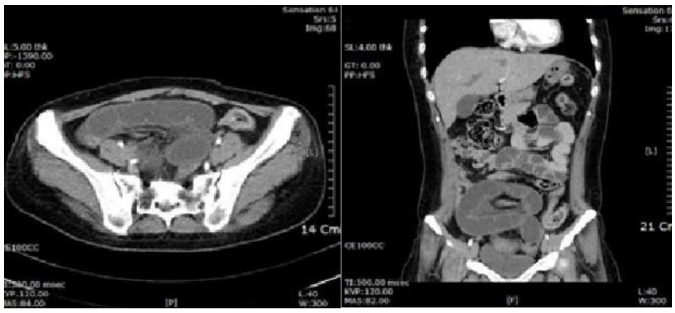

Figure 2: A computed tomography scan of abdomen demonstrating intestinal dilation with wall thickening.

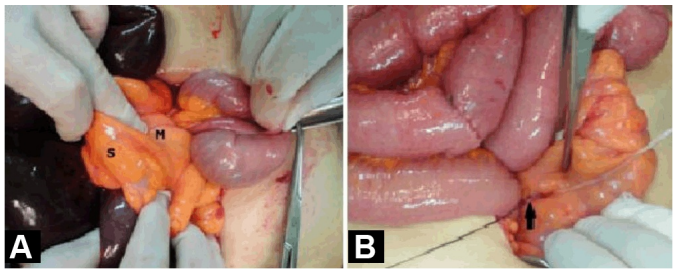

Figure 3: Operative findings (A) Herniation of small bowel through the sigmoid mesocolon. Herniated small bowel was necrotized. (S, sigmoid colon, M, sigmoid mesocolon), (B) Mesocolon defect next to the forcep (black arrow), which is oval and about three cm in diameter.

Intersigmoid Hernia: Herniation into an intersigmoid fossa, situated at the attachment of the lateral aspect of the sigmoid mesocolon.

Transmesosigmoid hernia: Incarceration of intestinal loops through an isolated, oval defect in the sigmoid mesocolon.

Intramesosigmoid hernia: A congenital, oval defect unrelated to the intersigmoid fossa is present in the lateral peritoneal surface of the mesocolon, and herniation occurs. A normal fusion fascia is present, and the right leaf is intact in this setting.

In our report, the patient had transmesosigmoid hernia according to the Benson classification. Pathologic apertures of the mesentery and visceral peritoneum are mostly due to congenital, surgical, traumatic, inflammatory or circulatory etiologies [2]. Congenital causes of sigmoid mesocolon hernias have also been proposed as possible causes [5–7]. However, the role of congenital factors remain obscure and theoretical. Some case reports have documented transmesosigmoid hernias developing during pregnancy or postpartum [5, 8]. The authors of these case reports proposed that dilatation and shrinkage of the uterus concomitant with pregnancy or delivery contributed to the development of transmesosigmoid hernias. The abnormal aperture could have been formed from the sigmoid mesocolon tearing by traction due to postpartum shrinkage of the enlarged uterus, and herniation could have occurred a few decades later through the abnormal aperture formed during the pregnant period. In our case, the patient had a laparotomy for tubal pregnancy 20 years prior, and delivery at 16 years prior. Her mesocolic defect was near the base of the sigmoid mesentery; operative trauma is therefore not suspected as the cause of the deep seated mesosigmoid defect. It could be a naturally occurring abnormal opening related to pregnancy. The key CT scan findings for diagnosis of the transmesosigmoid hernia included:

(a) a cluster of dilated fluid-filled loops of the small bowel entrapped in the left posterior and lateral aspect of the sigmoid colon through a mesosigmoid defect
(b) the defect located between the sigmoid colon and the left psoas muscle
(c) the sigmoid colon showing anterior and medial displacement
(d) these encapsulated loops of small bowel showing U or C shaped configurations and wall thickening representing closed loop obstruction and ischemic change
(e) attached mesentery with vessels engorgement and fat obliteration indicating strangulation and
(f) proximal small bowel showing dilatation [9].

Patients with small bowel obstruction not responding to conservative management requires operation. If an internal hernia is suspected, the operation should be prompt, as strangulation and gangrene of the bowel is likely to occur if the surgery is delayed. The role of laparoscopy in patients with intestinal obstruction is being increasingly recognized. Laparoscopic examination established that there was a strangulated hernia, and the definite etiology was only confirmed by the open method [10]. In contrast to other types of sigmoid mesocolon hernias, transmesosigmoid hernias are usually associated with a significantly long intestinal loop herniated into the opposite side of the mesocolon, perhaps because of a trend toward protrusion of the intestine due to the absence of the hernia sac [3, 4].

CONCLUSION

Small bowel obstruction secondary to transmesosigmoid hernia, although rare, may be considered in the differential diagnosis in patients with suspected small bowel incarceration or strangulation. We emphasize that in order to reduce morbidity and mortality from bowel strangulation, it is important to make the decision for surgical intervention by carefully watching the clinical features rather than identifying the precise cause of the acute abdomen.

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Author Contributions

Ji Won Kim – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Dae Hyun Yang – Conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Pulmonary renal syndrome in the Arabian Gulf region: A case report

Abdelkarim Waness

ABSTRACT

Introduction: Pulmonary renal syndrome refers to any pathologic condition that presents with varying concomitant lung and kidney symptoms and manifestations due to a common pathophysiologic insult. They are rarely encountered by health care professionals, including in the Arabian Gulf region, but can have severe therapeutic consequences. Case Report: We present a case of microscopic polyangiitis that manifested itself with pulmonary hemorrhage and acute kidney injury. The patient survived the initial insult but developed end-stage renal disease requiring ongoing hemodialysis. This report will review etiologies of pulmonary renal syndrome, clinical signs, diagnostic modalities, and therapeutic options for patients afflicted with this syndrome. Conclusion: Diagnostic and therapeutic challenges remain with pulmonary renal syndrome in the Arabian Gulf region despite advanced therapeutic interventions such as renal transplantation.

Keywords: Microscopic polyangeiitis, Alveolar hemorrhage, Plasmapheresis, Hemodialysis

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INTRODUCTION

Pulmonary renal syndrome (PRS) are rare medical syndrome defined by the co-existence of pulmonary hemorrhage and renal insult. PRS are, in fact, the clinical end-result many pathophysiologic entities. They can have variable presentations from dramatic respiratory distress to occult manifestations. PRS can represent both a diagnostic and therapeutic challenge requiring a close multidisciplinary teamwork. Cases of PRS are, occasionally, diagnosed in the Arabian Gulf region; namely within the Gulf Cooperation Council Countries (Saudi Arabia, Kuwait, United Arab Emirates, Qatar, Bahrain, Oman), Iraq and Iran.

CASE REPORT

A 60-year-old female, with history of diabetes mellitus, essential hypertension and hypothyroidism presented to a tertiary care center with dyspnea, cough and moderate hemoptysis for four days. Her medications included insulin, an ace inhibitor, and thyroid hormone replacement. On examination, she was in mild respiratory distress. Her vital signs were as follows: temperature 37.2°C, pulse 102 bpm, respiratory rate 18/minute, blood pressure 169/56 mmHg, and oxygen saturation 98% on room air. She had audible rhonchi especially in the left upper chest area. The rest of her examination was unremarkable. Laboratory results were (normal range in brackets): serum sodium 135 mmol/L (135–145 mmol), serum potassium 7.4 mmol/L (3.5–5.0 mmol), serum chlorine

106 mmol/L (98–107 mmol), bicarbonate 16 mmol/L (22–29 mmol), BUN 32.6 mmol/L (< 8.3 mmol), serum creatinine 657 µmol/L (it was 136 µmol/L seven months prior to this admission) (62–106 dL), WBC 9.4x10^3/mm^3 (4.5–11.0x10^3/mm^3), hemoglobin 8.2 g/µmol/L (12.6–17.4 g/L), platelets 193x10^3/µL (140–400/mm^3), INR 1.0 (0.8–1.2), PTT 26.8 (27.7–42.1). Her c-ANCA level was negative, the p-ANCA level was elevated at 79 U/mL (<5). Anti-nuclear antibodies (ANA), rheumatoid factor (RF), anti-DNA antibodies, anti-small muscle antibodies, cryoglobulin level, complements C3 and C4 levels were all normal. Chest X-ray showed diffuse patchy airspace opacities mostly in the left lung (Figure 1). Computed tomography (CT) scan of the chest confirmed extensive left lung ground glass opacities and interstitial thickening with patchy involvement of the right upper lobe (Figure 2). She underwent bronchoscopy with broncho-alveolar lavage (BAL) that visualized blood in the bronchioles, worse on the left side. Cultures for acid-fast bacilli (AFB), fungal and viral causes were negative. Because of deteriorating renal function, hemodialysis was initiated. Left kidney biopsy showed diffuse diabetic glomerulosclerosis, acute pauci-immune glomerulonephritis with mesangial cellular expansion, cresecentic formation and tubular epithelial cell changes (Figure 3). Her final diagnosis was microscopic polyangiitis. The patient was treated with corticosteroids and plasmapheresis. Her hemoptysis resolved slowly. However, she progressed to end-stage renal disease during her hospital stay. Six weeks post-discharge, the patient was clinically stable undergoing hemodialysis three times a week.

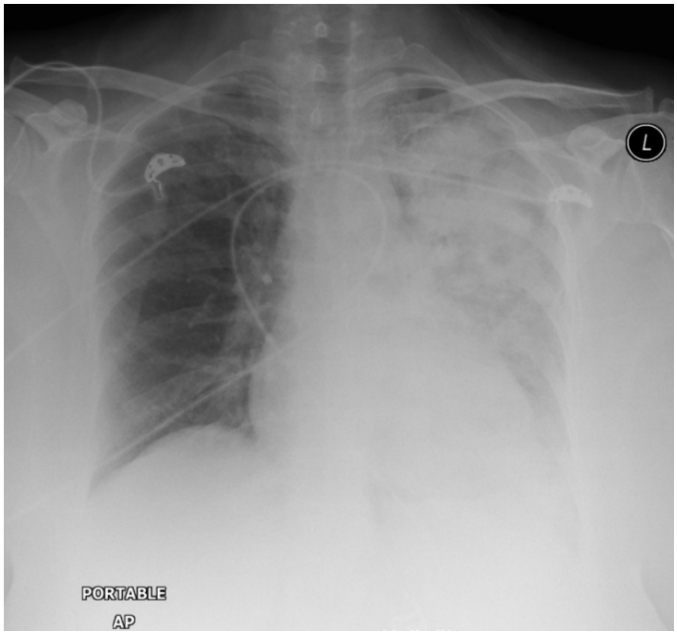

Figure 1: Chest X-ray with diffuse patchy airspace opacities more pronounced in the left lung.

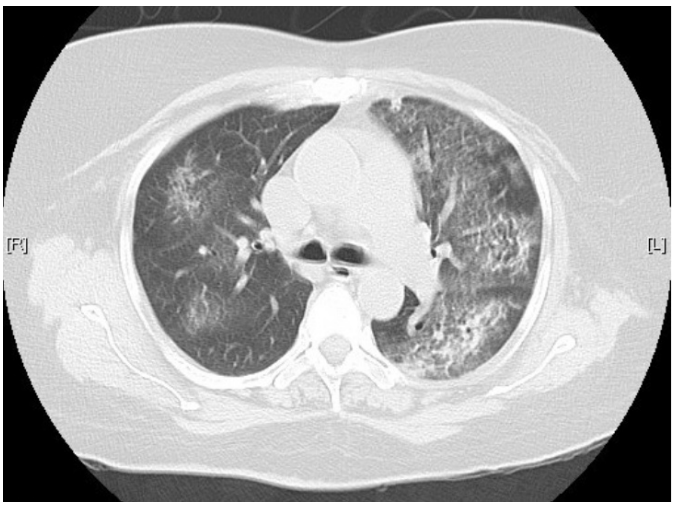

Figure 2: Computed tomography scan of the chest showing left lung ground glass opacity and interstitial thickening.

DISCUSSION

Causes of Pulmonary Renal Syndromes Goodpasture's syndrome

This was first described in 1919 by Ernest Goodpasture. This autoimmune condition can be interchangeably confused with PRS. The histopathological hallmark of this disease is the presence of anti-Glomerular basement membrane (anti-GBM) antibodies directed against type IV collagen in the GBM. Their immunological insult can affect both kidneys and lungs resulting usually in rapid progressive glomerulonephritis associated with alveolar hemorrhage. Occasionally, the insult is confined only to the kidneys or lungs separately. Goodpasture's syndrome, in association with primary systemic vasculitides, constitutes the most frequent etiologies of PRS. The prevalence of anti-GBM disease is low in the Arabian Gulf region (it is estimated between 1.5–4.4%) [1]). The diagnosis is suspected clinically and confirmed by renal biopsy. Prognosis is worse for patients who require immediate dialysis at presentation. Treatment armamentarium include immunosuppression, plasmapheresis, dialysis, and kidney transplantation.

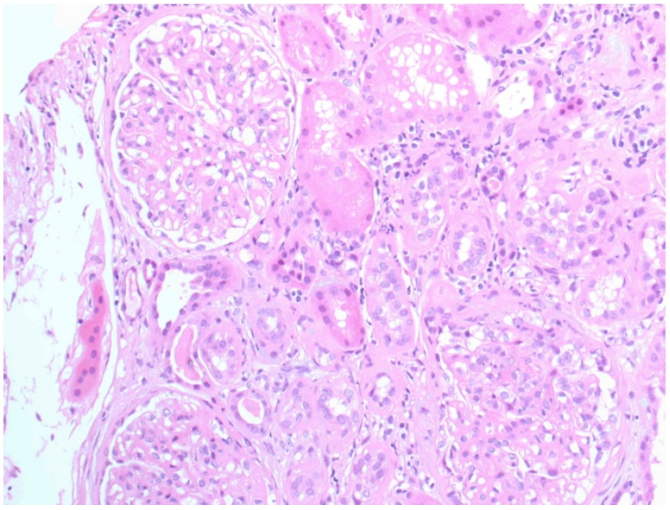
Figure 3: Kidney biopsy (Hematoxylin & Eosin stain) showing diffuse diabetic glomerulosclerosis, acute pauci-immune glomerulonephritis with mesangial cellular expansion, cresecentic formation and tubular epithelial cell changes.

Systemic vasculitides

These include different varieties of necrotizing conditions affecting large, medium, or small size vessels. Antineutrophil cytoplasmic autoantibodies (ANCA) seens to play a major pathophysiological role in many vasculiditides. Two principal serological subtypes of ANCA may be detected. Bosch et al. detected, in their study of 95 patients detected 26% and 74% cytoplasmic pattern (c-ANCA) and perinuclear pattern (p-ANCA), respectively. The majority of c-ANCA corresponded to anti-proteinase 3 (anti-PR3) antibodies (diagnostic for Wegener's granulomatosis); while the overwhelming p-ANCA corresponded to anti-myeloperoxidase (anti-MPO) antibodies principally detected in patients with rapidly progressive glomerulonephritis (RPGN) and hemorrhagic alveolar capillaritis [2]. Recent research have identified a new ANCA, directed against human lysosome membrane protein-2 (LAMP-2), that possibly has a concurrent action with PR3-ANCA or MPO-ANCA, for renal ANCA-associated vasculitides (AAV) [3]. The presence of ANCA and their subsequent histological immune complex deposits is not a prerequisite for the development of necrotizing arteritis. Primary systemic vasculitides are an important cause of PRS. Occasionally, cases of ANCA-positive Goodpasture's overlap syndrome are diagnosed.

Wegener's granulomatosis: Can share pathogenic, pathological, and clinical features with other vasculiditides. This necrotizing arteritis has predilection for the upper respiratory system, but can cause hemoptysis and glomerulonephritis. Sporadic cases of Wegener's granulomatosis causing PRS are rarely discovered [4].

Churg-Strauss syndrome (CSS): First described in 1951 and also known as 'allergic granulomatous angiitis', CSS is another ANCA-associated arteritis. Asthma is a central feature of this condition. Although less frequent, alveolar hemorrhage and renal involvement can be observed. Adult and pediatric cases are occasionally diagnosed in the Gulf-countries [5].

Microscopic Polyangiitis (MPA): The most common ANCA-associated small vessel vasculitis. Most patients will exhibit MPO-ANCA positive serology; however PR3-ANCA serology can be present [6]. It can affect any body organ, pulmonary hemorrhage and nephritis are common. The latter usually progresses to end-stage renal disease requiring continuous hemodialysis.

Polyarteritis Nodosa (PAN): A systemic vasculitis affecting medium and small muscular arteries, preferentially at vessel bifurcations, resulting in microaneurysm formation, aneurysmal rupture with hemorrhage, thrombosis, and, consequently, organ ischemia or infarction ANCA is rarely positive in PAN; with positive MPO-ANCA however, PAN can have close clinical resemblance to MPA [7].

Behçet's syndrome (BD): A multisystem disease of unknown etiology with well-established presence in the Gulf countries BD has many symptoms including neuropsychiatric expressions, oral and genital ulcerations up to 5-10% of patients with this condition can have pulmonary manifestations such as hemoptysis [8].

Henoch-Schonlein Purpura (HSP): Most common systemic vasculitis in children with documented cases in the Gulf countries [9]. It is usually a self-limiting disease. Pulmonary complications, such as pulmonary hemorrhage, can be observed in HSP [10].

Mixed Cryoglobulinemias: Vasculitides with close association to some connective tissue diseases such as systemic lupus erythematosis (SLE), and Sjogren syndrome. Viral infections, particularly hepatitis C infection, play a role in the development of this pathology. Hepatitis C infection is prevalent in the Gulf region especially among patients undergoing chronic hemodialysis, and cases of cryoglobulinemias are routinely encountered [11].

Renal disorders

Immunoglobulin A (IgA) nephropathy: First described by Berger and Hinglais in 1968. IgA nephropathy is characterized by predominant IgA deposition in the glomerular mesangium. Its prevalence in the Gulf countries has been increasing recently according to a study from Bahrain by Arrayed et al. [12]. Gross hematuria and renal insufficiency are the main features of IgA nephropathy, however it can rarely cause alveolar hemorrhage [13]. The mechanism of PRS in IgA nephropathy is probably linked to a pauci-immune vasculitis induced by IgA ANCA [14]. Diagnosis is confirmed by kidney biopsy. Prognosis of IgA nephropathy is generally favorable although some cases progress to end-stage renal disease

Idiopathic immune-complex glomerulonephritis (IICG): Can be a misnomer since it is not confined to the kidneys; indeed, cases of IICG with alveolar hemorrhage have been documented. Furthermore, it is suggested that both anti-MPO antibodies and serum MPO are closely related to the pathogenesis of idiopathic crescentic glomerulonephritis [15].

Connective tissue disorders

Systemic lupus erythematosis (SLE): Prevalent in the Gulf countries. It can affect both genders and different age groups [16]. Lupus nephritis is a very common complication occurring in 37–69% of patients affected with SLE in a study from the United Arab Emirates [17]. Furthermore, SLE can have a wide spectrum of other manifestations including intractable alveolar hemorrhage.

Rheumatoid Arthritis (RA): Routinely encountered in the Gulf region since decades agoA recent Saudi study reported a high mortality rate of 16% in local population having extra-articular manifestations of RA including renal and respiratory tracts involvement [18].

Progressive Systemic Sclerosis (PSS): The genetic makeup of the local population may play a role in the development of such condition [19]. Rare cases of PSS or mixed connective tissue disease (MCTD) associated with MPO-ANCA-positive arteritis can be diagnosed. They cause crescentic glomerulonephritis and hemoptysis [20].

Sjogren's syndrome (SS): Also known as sicca syndrome, is a chronic auto-immune inflammatory disease affecting primarily the lacrimal and salivary glands occasionally observed in the Gulf countries. Although rare, it can cause glomerulonephritis. On the other hand, pulmonary manifestations and particularly non-specific interstitial pneumonia bullae formation, and pulmonary nodular amyloidosis. SS causing diffuse pulmonary hemorrhage and subsequently PRS has been documented [21].

Polymyositis/Dermatomyositis: An idiopathic inflammatory myopathy known with its possible malignancy association. Among other symptoms, it can cause severe diffuse alveolar hemorrhage. Sporadic cases are documented in the region [22].

Other etiologies of PRS: Other potential etiologies that might be observed in the Gulf area include:

Anti-phospholipid syndrome (APS): An auto-immune disorder characterized by the wide-spread development of both arterial and venous thrombosis. APS was documented to cause PRS rarely [23].

Drugs: Such as hydralazine, allopurinol, sulfasalazine, or penicillamine can induce ANCA-positive vasculitis or anti-GBM-like disease and subsequently manifest itself in PRS [24].

Occupational exposure: A relationship between ANCA-associated (especially MPO ANCA)-glomerulonephritis and silica exposure has been postulated with documented cases of PRS [25].

Signs and symptoms: The two main clinical pillars for pulmonary renal syndromes include: pulmonary symptoms and renal symptoms. Pulmonary hemorrhage can have spectacular and dramatic presentation, and patients can present with cough, wheezing, dyspnea and possibly cyanosis. Renal symptoms can be subtle or manifest such as flank pain, hematuria, oliguria and renal insufficiency. Constitutional symptoms such as fever, chills, fatigue, arthralgia, myalgia, and weight loss are common. A variety of specific symptoms related to the underlying medical condition can be seen. Seizure, migratory mononeuritis multiplex, or even stroke can be the vasculitic manifestations within the nervous system. Oral ulcerations and even gross nasal septum or palatal destruction are prominent with WG and BD. Leukocytoklastic (palpable) purpura and even cutaneous ulceration can be a prominent feature with HSP and PAN. Connective tissue conditions can offer a wealth of clinical signs such as shiny tight skin, cutaneous telangiectasias or calcinosis, malar or heliotrope rashes, arthritic deformities; or signs and symptoms of pleural, pericardial, or peritoneal irritation. Clinicians must be on the look-out for such findings in order to diagnose and treat the underlying condition, and not only the PRS manifestations.

Diagnostic tools: Many laboratory findings in PRS are non-specific such as anemia, leukopenia, thrombocytopenia, elevated sedimentation rate, increased serum creatinine level, electrolyte disturbances, and presence of red blood cells (RBC) or RBC-casts in the urine. More specific laboratory tests are dictated by the clinical picture. These include antinuclear antibodies, anti-double strand DNA antibodies, rheumatoid factor, anti-neutrophil cytoplasmic autoantibodies (ANCA), anti-phosphlipid antibodies, anti-smooth muscle antibodies, cryoglobulin level. Radiologically, chest X-ray can be normal or show alveolar infiltrates, pulmonary nodules or cavitations. CT scan of the chest can confirm prior observations or discover occult benign or malignant findings. Renal sonography can be normal or possibly shows stigmata of ongoing prior kidney disease. Bronchoscopy with BAL and possibly tissue biopsy are very valuable. The diagnosis of PRS is clinched after doing a kidney biopsy in the majority of cases. Other tissue samples, such skin or nerve biopsies, might help clarify the diagnosis. If the co-existence of pulmonary hemorrhage and renal injury is purely coincidental and has no immunopathologic relationship, the condition then is termed *false-positive PRS* or *non-specific PRS* [26].

PRS treatment considerations: The therapeutic approach to PRS can be complex and is generally multidisciplinary. Some considerations in the management of PRS are:

(a) *Educational:* Consanguinity is prevalent in the Gulf countries. It would be of great benefit to educate the general population about the risks of such practice. Further, stressing patients' education about PRS and its possible complications is paramount.

(b) *Supportive therapy:* This must be provided in earnest within the confinement of qualified centers. Supportive care can be vital in the management of PRS. Indeed, it has been proven that measures such as temporary mechanical ventilation and hemodialysis can reduce patients' mortality [27]. Moreover, physical and psychological supports might be needed for some patients.

(c) *Immunosuppression*: Corticosteroids and immunosuppressors, such as cyclophosphamide aim at cutting down the production of auto-antibodies seen in many PRS [28]. Plasmapheresis assists with the removal of already circulating auto-antibodies. Immuno-suppression comes, however, with a heavy price at times such as drug toxicity and decreased immunity resulting in severe infections.

(d) *Dialysis:* Patients with PRS usually develop rapid progressive form of glomerulonephritis requiring early dialysis intervention. Despites appropriate support, the majority of them will lose the remaining kidney function after the initial insult. Such patients will end up dialysis-dependant for a prolonged period of time [29].

(e) *Renal transplantation*: This is another option for patients afflicted with auto-immune conditions complicated by end-stage renal disease. In the Gulf countries, and in spite of recent improvement, there is still an increasing demand for kidney transplantation done both locally and abroad [30].

(f) *Medical Research:* Clinical research, such as by the National Institutes of Health in the United States of America, is being conducted to find possible genetic factors in the pathogenesis of some of the etiologies of PRS. Despite generous financial resources in the most Gulf countries, medical research remains restricted with limited funding.

CONCLUSION

Pulmonary renal syndrome occur routinely in the populations of the Arabian Gulf. All possible etiologies are encountered with similar aggressiveness as other regions of the world. Despite major leaps in healthcare improvement, ongoing challenges such with renal transplantation persist. A better medical research pathway for such pathologies certainly needs to be boosted in this region.

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Author Contributions

Abdelkarim Waness – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Massive and sustain potassium therapy saves life in barium chloride intoxication: A case report

Mohy Kadri El Masry, Walaa Gomaa Abdelhamid, Salma Ibrahim Abdelkader, Sara Ahmad Elmorsi, Sara Atef Abdelaziz

ABSTRACT

Introduction: Barium salts present real threat to patients if the radiocontrast material contains soluble barium contaminant as the chloride form which is one of the most soluble salts. Case Report: A patient 37-year-old was given 40 g barium chloride as radiocontrast for gastro-esophageal reflux imaging. It was immediately followed with severe vomiting and hematemesis. Shock, ventricular tachycardia, severe flaccid paralysis, coma and severe hypokalemia followed within the first hour. The patient responded favorably to large infusion of IV potassium, reaching 40 mmol hourly for the first three hours. Potassium infusion was continued for the first four days of treatment. A total IV potassium infusion of 560 mmol was given in the first day. Abundant IV fluids infusion contributed to the correction of severe dehydration caused by vomiting and diarrhea. Prerenal failure and ischemic hepatitis secondary to shock improved with hemodynamic correction. Conclusion: Following barium poisoning, severe hypokalemia (1.2 mmol/L in this case) is responsible for ventricular tachycardia, shock, flaccid paralysis and respiratory failure. Large IV potassium infusion should continue for the first few days and is considered the principal therapeutic guideline in barium poisoning and the mainstay for the correction of almost all vital functions.

Keywords: Barium poisoning, Hypokalemia, Ventricular tachycardia, Paralytic respiratory failure

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El Masry MK, Abdelhamid WG, Abdelkader SI, Elmorsi SA, Abdelaziz SA. Massive and sustain potassium therapy saves life in barium chloride intoxication: A case report. International Journal of Case Reports and Images 2012;3(12):35–38.

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INTRODUCTION

Insoluble barium sulfate is used in its pure form for the insolubility, as an X-ray radiocontrast agent for imaging the human gastrointestinal tract. Rare cases of unintentional toxicity have been reported during radiographic procedures and include complications associated with oral administration [1] of soluble barium salts unintentionally contaminating contrast solution [2]. Soluble barium compounds like acetate, chloride, hydroxide, nitrate are poisonous due to release of the soluble barium ion, and have been used as rodenticides. The solubility of barium chloride is high and typically produces a rapid onset of life threatening

manifestations. In addition to good supportive care, the mainstay of treatment is rapid correction of hypokalemia [3].

CASE REPORT

A 37-year-old male was transferred by ambulance from a district hospital following uncompleted barium meal radioimaging procedure, during which the patient developed upper abdominal pain, recurrent vomiting, hematemesis and profuse sweating followed by hypotension, tachycardia and drowsiness. Present history revealed that the patient was requested to supply the barium for the imaging as the hospital was short of the chemical. Radiology technician realized that he gave the patient a suspension containing the equivalent of 40 g barium chloride instead of barium sulfate when he revised the container. Serum potassium was 2.2 mmol/L one hour after the accident. He was immediately given 5 g oral magnesium sulfate, 40 mmol KCl in IV fluids, 200 mg hydrocortisone and metoclopramide.

On arrival to poison control center (PCC), as evident in Table 1 he was in deep flaccid coma grade IV (Reeds classification); he had occasional fasciculations of small muscles of hands and face. Pupils were dilated and sluggishly reacting. He was in shock with systolic blood pressure of 50 mmHg, diastolic blood pressure of 30 mmHg, and barely palpable peripheral pulsations. ECG showed ventricular tachycardia.

Table 1: Progressive clinical and laboratory notes in response to advocated management

	1-2 h	PCC 3rd h	4th h	6th h	8th h	12th h	24th h	Day 2	Day 3	Day 4	Day 5
Blood Pressure (mmHg)		50/20	90/60	150/100		130/80		120/80			
ECG		VT - VF		ST- PVCs		VT		PVCs		Sinus	
Vomiting (mL)	700	300			200	200		200	50		
Diarrhea (mL)		800					500	500			
Coma (Grade)		III/IV		III			I	0			
Muscle Power		0			1		3	4	5		
PO2 mmHg		31.1	69.3					227.5	159	80.9	137.7
PCO2 mmHg		58.3	42					38.7	36.5	38	43.9
PH		7.29	7.26					7.31	7.43	7.48	7.46
SaO2		51	91.3					99.6	99.5	96.4	99.0
Serum potassium (mmol/L)	2.2	1.2	2.4		2.5	1.8	4.0	3.7	3.6	3.6	3.6
Hematocrit		55.7						54.5			51%
WBCs x (10^3/mm^3)		26.6						23.5			11
SGPT (IU/L)			37					105		97	73
SGOT (IU/L)			42					148		161	109
Urea (mg/dL			83					65	47	41	
Creatinin (mg/dL)			1.8					1.6	1.3	1.1	
Gastric lavage		+			+						
Dose KCl (mmol)	40	40	40	40	40	120	240	200	120	80	
Antiarrhythmic		Lidocaine 100 mg bolus followed by 2 mg/min			Amiodarone 150 mg IV over 20 min, then 1 mg/min (6 h) then 0.5 mg/min						
MgSO4 (gm)	5 gm	15			15						
Dopamine			5 µ/kg/min		Weaned						
IV Fluids (ml)		1500	1000	1000		6450		3400	2250	2750	
Input (+)/ Output (-)			+2150			+2950		+900	-750	- 500	
Mechanical Ventilation				Controlled				SIMV	Wean T tube	Extubat i-on	

Abbreviations: h - hour, VT - Ventricular tachycardia, VF - Ventricular fibrillations, PVC - Premature ventricular contractions, SIMV: Synchronized intermittent mandatory ventilation

Immediate cardiopulmonary resuscitaion (CPR), endotracheal intubation and respiratory support were undertaken. Initial assessment revealed respiratory failure with PaO2 31.1 mmHg, PaCO2 58.3 mmHg; SaO2 51% and serum K 1.2 mmol/L. Intravenous fluids with large doses of potassium were immediately infused with a high potassium rate reaching 40 mmol hourly in the first 3 hours and 20–30 mmol/L hourly thereafter for the first day guided by serum potassium levels. Gastric lavage with abundant tap water was performed followed by instillation of 10 g magnesium sulfate. This was repeated after three hours. Blood pressure was rapidly corrected to 90/60 mmHg in response to large IV fluid administration, and antiarrhythmic therapy.

A gradual regain of consciousness and muscle power of hands and arms was noticed after the first 12 hours of treatment and spontaneous breathing was detected during mechanical ventilation. Lidocaine used initially was discontinued and followed by amiodarone. Both were given in recommended doses. Abundant IV fluids were followed by the correction of hemodynamic status. Dopamine started initially was rapidly weaned.

Serum potassium showed fluctuations between 1.8 and 2.5 mmol/L during the first 24 hours despite large potassium infusion rates. Patient receiving 560 mmol potassium on the first day. On the second day, 200 mmol potassium, and almost 120 mmol daily thereafter were required to keep serum potassium levels within low normal values. Starting from the second day serum potassium varied between 3.6 and 4.0 mmol/L.

Patient was initially on controlled mode mechanical ventilation and was shifted to SIMV mode on day 3, weaned and extubated on day 4.

Hematemesis and vomiting continued for four days despite high doses of parenteral omeprazole, vitamin K, tranexamic acid and cold water gastric lavage.

On the first day, hematocrit was 55.7%, total proteins 9 g/dL and total leucocytic count $26x10^3/mm^3$. A mild rise of urea and creatinine was evident on the first day and decreased slightly on the second day. CPK was normal. Liver transaminases were normal on admission but rose to 105 IU/L for SGPT and 148 IU/L for SGOP to gradually decrease daily thereafter. Neurological examination was normal before discharge on day 5.

DISCUSSION

Life threatening hypokalemia is uncommon in clinical toxicology practice. Moderate to severe hypokalemia are common after deliberate insulin overdose and theophylline toxicity. In barium toxicity, potassium redistribution and sequestration occur dramatically and rapidly inside the cells. Barium reduces potassium efflux from muscle cells by blocking potassium channels. Therefore, the continued activity of the ion pump combined with blocked potassium efflux results in extracellular hypokalemia and intracellular potassium accumulation. The serum potassium level can drop quite precipitously, and so does the trans-membrane ionic diffusion potential, to less than 60 mV. At this membrane potential the muscle is unexcitable and paralysis ensues [4].

The precipitous hypokalemia occurring in our case, reaching 1.2 mmol/L, was evidently responsible for the appearance of paralytic respiratory failure, ventricular tachycardia and episodes of ventricular fibrillation and eventually shock.

Despite the rapid CPR, lidocaine infusion and mechanical ventilation, it was evident that the concomitant intravenous infusion of large doses of potassium chloride reaching 120 mmol in the first three hours, as a combination treatment, was responsible for the correction of rhythm disturbances and shock state. This was previously confirmed and recommended [5, 6]. Despite the high rate of potassium infusion, we estimate the rate of potassium infusion was slower than the ideal. Under lidocaine infusion and later amiodarone recommended dosages treatment, recurrence of non-sustained ventricular tachycardia and premature ventricular ectopy, was strongly correlated with slowing rate of potassium infusion to 20 mmol/h and relapsing life-threatening hypokalemia (1.8 mmol/L) necessitating the resumption of massive infusion of potassium greater than the dosage recommended by some authors although encouraged by others [5, 7]. This demonstrates that blocking barium effect on the potassium rectified channels and block of potassium efflux continued till the 12^{th} hour after ingestion. With potassium infusion reaching 560 mmol in the first 24 hours, serum potassium still tended to decline after initial correction to 1.8 mmol/L

A non-negligible contributing effect to hypotension and shock is the dehydration provoked by vomiting and third space fluid entrapment in the digestive tract related to barium and magnesium sulfate. Large infusion rates with positive fluid balance (+6100 mL on the first day) in our case corrected the laboratory signs of dehydration evidenced by an elevated total proteins (90 g/L) and hematocrit (55.4%) which returned to 51% on discharge. This case highlights the importance of dehydration in barium poisoning in precipitation of shock.

Ischemic hepatitis secondary to shock was responsible for the mild rise of transaminases on day 2. Elevated serum transaminases gradually returned to normal after correction of blood pressure and control of shock. Severe blood tinged vomiting persisting till day 4 of poisoning confirming hemorrhagic gastritis and esophagitis for which H2 blockers and proton pump inhibitors were essential part of our treatment strategy.

Peripheral respiratory failure secondary to serious flaccid paralysis and ultimately respiratory muscle weakness in our case, necessitated immediate and prolonged control mode mechanical ventilation. The gradual regain of muscle power by the end of the first day closely coincided with the correction of severe hypokalemia, and permitted the initiation of SIMV mode ventilation. This observation indicated the correlation of profound muscle weakness to hypokalemia rather to the barium itself.

This case represents the largest barium chloride dose (40 g) required to survive poisoning and highest IV

potassium requirements (560 mmol in the first 24 hours) so far reported in literature.

The case presents a serious form of medical malpractice related to administration of barium chloride, non-verified by the radiology technician before using for radiocontrast imaging. This life-threatening situation is caused by the lack of clear strategic guidelines for checking the safety of this procedure, deficient provision of drugs and contrast materials in hospitals and dependence on patients to purchase the materials and drugs required to accomplish the due care.

CONCLUSION

The critical communication conveyed by this case of acute barium poisoning, is the importance of early and massive potassium administration in doses even higher than recommended in literature to correct severe life-threatening recurrent ventricular arrhythmias, shock and paralytic respiratory failure. Even with toxic doses of soluble barium compounds higher than described in literature, survival is possible with early, aggressive and sustained proper management. Dehydration and ischemic hepatitis concomitant with shock, although rarely described, might complicate acute barium poisoning.

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Author Contributions

Mohy Kadri El Masry – Conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Walaa Gomaa Abdelhamid – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
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Sara Ahmad Elmorsi – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Sara Atef Abdelaziz – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Recurrent opportunistic infections in a post-transplant lymphoma patient

Ayelet Raz-Pasteur, Yaakov Dickstein, Ilana Oren, Ayelet Eran, Irit Avivi, Khetam Hussein

ABSTRACT

Introduction: Infections and graft-versus-host disease (GVHD) are the most notable problems after allogeneic stem cell transplantation (Allo SCT). Case Report: A 50-year-old patient underwent an Allo SCT for transformed follicular lymphoma (t-FL). Transplant course of the patient was complicated by GVHD and recurrent, severe, opportunistic infections including pneumonia caused by two fungal pathogens simultaneously, recurrent bacteremia with no apparent source, septic arthritis caused by an unusual pathogen and eventually developed progressive multifocal leukoencephalopathy (PML). Conclusion: This case illustrates the diversity of opportunistic infections affecting patients undergoing Allo SCT and the complexity involved in diagnosis and treatment of such infections.

Keywords: Allo SCT, GVHD, Opportunistic infections, Follicular lymphoma (FL), PML

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INTRODUCTION

Infections and graft-versus-host disease (GVHD) are the most notable problems after allogeneic stem cell transplantation (Allo SCT).

We describe a 50-year-old patient who underwent an Allo SCT for transformed follicular lymphoma (t-FL) and whose transplant course was complicated by GVHD and recurrent, severe, opportunistic infections including pneumonia caused by two fungal pathogens simultaneously, recurrent bacteremia with no apparent source, septic arthritis caused by an unusual pathogen and eventually developed progressive multifocal leukoencephalopathy (PML).

CASE REPORT

A 50-year-old male patient was diagnosed with t-FL and underwent an Allo SCT from matched sibling donor without T-cell depletion, resulting in durable complete remission. His posttransplantation course was initially complicated by an early onset of acute GVHD, grade III, with gastrointestinal (GI) and hepatic manifestations including diarrhea and deranged liver function tests (LFTs) including increased bilirubin which were successfully controlled with prednisone and cyclosporine. Prednisone was given at a dosage of 80 mg for several weeks.

One month later, the patient was hospitalized with high fever, general weakness, dry cough and dyspnea. At the time of admission, his vital signs were stable. Blood pressure was 125/70 mmHg and pulse was 86 bpm. Initial laboratory investigations including liver function tests were within normal range apart from macrocytic anemia hemoglobin (Hb) 9.5 g/dL and mild thrombocytopenia $7.2x10^5/mm^3$ with a normal neutrophil count. A chest X-ray (CXR) revealed bilateral lung infiltrates. On a chest computed tomography (CT) scan multiple nodular lesions with halo sign were noted. Urine Legionella antigen was positive and culture of bronchoa/vedas lavage (BAL) fluid grew *Legionella* species. *Aspergillus* DNA and *Legionella* DNA were detected in BAL fluid using polymerase chain reaction (PCR) and Galactomannan was negative, establishing the diagnosis of simultaneous *Legionella* pneumonia and invasive pulmonary aspergillosis (IPA). The patient was treated with voriconazole and levofloxacin and showed clinical improvement. The patient was on weekly follow-up with the hematology clinic and his condition was stable.

Three months Allo SCT, a multidrug-resistant *Aspergillus Campylobacter jejuni* was isolated from blood cultures drawn from a central venous catheter (CVC) and from peripheral blood as well as stool culture. The patient was treated with imipenem for two weeks. Extensive search for the source of recurrent bacteremia including venous ultrasound doppler examination and a trans-esophageal ecocardiogram (TEE) was unrevealing.

Concomitantly, the patient became disoriented. Brain CT scan and magnetic resonance imaging (MRI) were within normal limits. Lumbar puncture (LP) was performed, to obtain cerebrospinal fluid (CSF) for culture and molecular studies, for the detection of viral, bacterial and fungal pathogens. A laboratory results for viral, bacterial and fungal studies were negative. The neurologic manifestations were attributed to side effects of imipenem, and treatment was switched to meropenem. The patient gradually regained his previous cognitive status.

One month later, the patient was readmitted due to aggravation of left shoulder pain without fever. Ultrasound, CT and MRI of the shoulder were suspicious for septic arthritis (Figure 1). Arthrocentesis of the shoulder joint effusion revealed purulent fluid with white blood count of $4.1x10^4/mm^3$. Synovial fluid gram stain yielded gram positive bacilli and culture grew *Nocardia* spp. that were identified as *N. cyrageorgica* by PCR. Due to trimethoprim/sulfamethoxazole allergy, treatment with imipenem and amikacin was initiated.

Seven months after Allo SCT, the patient developed gait imbalance and delirium. Initial work-up revealed no electrolyte imbalances as well as normal renal function and LFTs. MRI demonstrated a diffuse process involving both frontal lobes (Figure 2). Lumbar Puncture was repeated. The CSF contained leucocytes $3/mm^3$, a normal glucose level, and slightly elevated protein level. Gram stain, CSF culture and cryptococcal antigen test were all negative. PCR of the CSF for

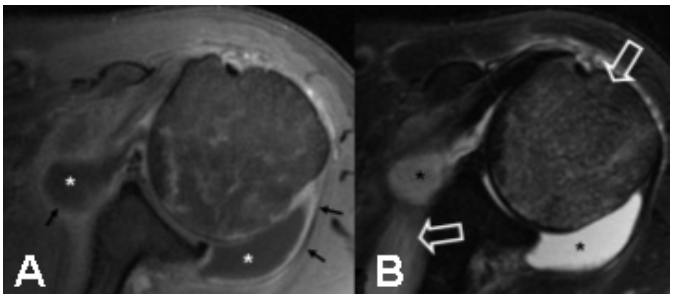

Figure 1: Axial T1 weighted image following gadolinium injection, (A) and axial T2 weighted, (B) MRI of the left shoulder. A large joint effusion (asterix) with peripheral enhancement (arrow) is noted. Additionally, edema of the humeral head and adjacent soft tissues is shown (B, open arrow).

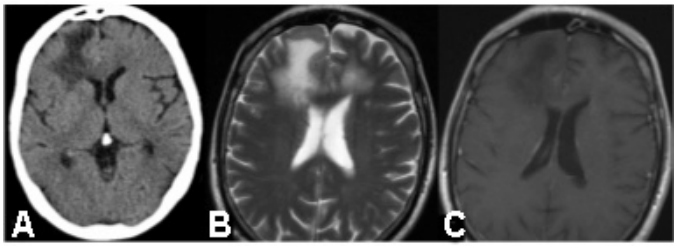

Figure 2: Axial CT, (A), and MRI images of the brain, T2 weighted, (B) and T1 weighted following gadolinium injection, (C). A white matter lesion is noted in the right frontal lobe on CT and bilaterally on MRI, right greater the left. The lesion extends to the subcortical white matter, does not enhance and has a mild mass effect (imaging characteristics typical for progressive multifocal leukoencephalopathy).

Epstein-Barr virus, Cytomegalovirus, Herpes simplex virus, human Herpes virus 6 and Varicella Zoster virus were negative. Serum serology for West Nile virus was negative. PCR for JC virus demonstrated more than a million copies/mL in CSF. An effort to reduce immunosuppression resulted in a flare of the GVHD. Treatment with mefloquine and liposomal cidofovir was initiated but the patient's neurological status continued to deteriorate until his death two weeks later.

DISCUSSION

Three risk periods of immunologic deficiency occur predictably in recipients of Allo SCT—the pre-engraftment period, the early post-engraftment period (until day 100) and the late period (after day 100).

The late posttransplantation period is heralded by the recovery of cell-mediated immunity (CMI) and humoral immunity. This phase begins at day 100 and continues until the bone marrow transplant (BMT) recipient stops all immunosuppressive medication for GVHD. During this period, viruses are responsible for more than 40%, bacteria for approximately 33% and fungi cause approximately 20% infections [1]. Our patient experienced recurrent infections, including

Legionella pneumonia and pulmonary aspergillosis followed by septic arthritis due to *Nocardia* and eventually PML caused by JC virus. Among these, IPA exhibits a bimodal distribution, with the first peak at two to three weeks posttransplant and the second peak at three to four months posttransplant, usually in conjunction with persisting GVHD. The remaining pathogens occur primarily in conjunction with immunosuppression connected to treatment of GVHD and appear more remotely after Allo SCT.

The initial CXR and CT in our patient suggested an atypical or opportunistic infection with difficulty in differentiating between IPA and *Legionella*. Immunosuppression increases the risk for invasive fungal infection and is consistently implicated as a risk factor for *Legionella* pneumonia with transplantation recipients at the highest risk [2]. Thus the need for BAL testing. Isolation of Legionella in the BAL in conjunction with positive Legionella PCR from BAL and positive urine *Legionella* Antigen, definitively verified the diagnosis of *Legionella* pneumonia in our patient.

As stated, the onset of *Aspergillus* infection after Allo SCT occurs in a bimodal distribution. In a recent epidemiological multi-center survey conducted between 1999 and 2003, aspergillosis was the most frequent fungal complication among patients receiving allogeneic transplant, among whom aspergillosis is responsible for 81% of all fungal infections [3]. The lungs represent the most frequently involved site for invasive aspergillosis and predisposing factors include powerful immunosuppressive chemotherapy, neutropenia and synergistic combinations of potent broad-spectrum antibiotics [4]. In our patient, specific risk factors included Allo BMT, immunosuppressive therapy, GVHD and occasional neutropenia.

As opposed to the diagnosis of *Legionella* pneumonia our patient did not meet the criteria of proven invasive pulmonary aspergillosis. In the case of suspected IPA, the evaluation should include a radiologic examination of the lungs, sinuses and brain, bronchoscopy and BAL for microscopy, culture and PCR and a test for circulating galactomannan [4]. The definite diagnosis of IPA requires a positive culture or histopathology. Our patient was diagnosed with IPA following a typical chest CT scan and the presence of positive PCR for *Aspergillus* in BAL, but BAL galactomannan antigen and culture for *Aspergillus* were notably negative.

The respiratory infection in our patient was followed by persistent *Campylobacter bacteremia* arising from his gut. Blood stream infection, in general, has been examined primarily in immunocompromised patients with neutropenic fever among whom approximately 50% are ultimately diagnosed with an infectious cause and 20% yield positive blood cultures. *Campylobacter* bacteremia is a rare disease, occurring mainly in patients with immune deficiency or other serious underlying conditions [5]. In an unpublished study conducted in our i nstitution, *Campylobacter* bacteremia occurred chiefly among severely immunocompromised patients, especially those with hematological malignancy. *Campylobacter jejuni* subsp. *Jejuni* is the main bacterial cause of enteroinvasive diarrhea, and it is rarely complicated by bacteremia or extraintestinal localization [6].

Shortly after the bacteremia, our patient presented with septic arthritis caused by *Nocardia*. Immunodeficiency is a well-established risk factor for nocardiosis. Pulmonary disease is the predominant clinical finding of systemic nocardial infection, with the CNS as the second-most involved system in one large survey [7]. Nocardial septic arthritis is significantly less common, and is usually attributed to hematogenous spread in immunocompromised hosts [8]. *N. asteroides* complex is the most common species causing septic arthritis, but *N. braziliensis*, *N. caviae* and *N. farcinica* have also been identified.

The last infectious complication which developed in our patient was PML. As previously described, the patient presented with neurologic, clinical and radiologic findings suggestive of pyogenic brain abscess, which in the immunosuppressed patient has a broad differential diagnosis including viral, bacterial and fungal agents. Amongst these, the most common ubfectuibs agents include the herpes virus group and adenovirus, infection with the bacteria *Listeria* and *N. asteroides*, *M. tuberculosis*, *Mucorales* sp, *Candida* and *C. neoformans* [9].

Our patient was diagnosed with PML. This rare and usually fatal viral disease is characterized by progressive damage or inflammation of the white matter of the brain at multiple locations. It occurs almost exclusively in people with severe immune deficiency and is caused by JC virus, which is normally present and kept under control by the immune system. Immunosuppressive drugs prevent the immune system from controlling the virus and reactivation can occur. Symptoms include weakness or paralysis, vision loss, impaired speech and cognitive deterioration.

PML is diagnosed by the finding of JC virus DNA in CSF or brain biopsy. Characteristic evidence of the damage caused by PML in the brain can also be detected on MRI images which classically show multifocal non-enhancing lesions without mass effect.

Currently, there are no approved therapies for PML. Although a number of preclinical reports and case studies have suggested the potential anti-PML effects of antiviral and antineoplastic drugs such as cytarabine, cidofovir and topotecan, larger case-controlled studies failed to establish the efficacies of these medications. *In-vitro* data coupled with biodistribution data suggest that mefloquine could represent an effective therapeutic agent for the treatment of PML [10]. Accordingly, our patient was treated with the combination of mefloquine and cidofovir.

CONCLUSION

Our case illustrates that a range of opportunistic infections can affect a patient undergoing Allo SCT during a relatively short period of time. Furthermore,

the appearance of Nocardial infection in the form of septic arthritis is a rare presentation of this pathogen.

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Author Contributions

Ayelet Raz-Pasteur – Conception and design, Acquisition of data, analysis and interpretation of data, drafting the article, Final approval of the version to be published
Yaakov Dickstein – Conception and design, Acquisition of data, Critical revision of the article, Final approval of the version to be published
Ilana Oren – Conception and design, analysis and interpretation of data, Critical revision of the article, Final approval of the article
Ayelet Eran – Analysis and interpretation of data; Drafting the article; Final approval of the article
Irit Avivi – Analysis and interpretation of data, Drafting the article, Final approval of the article
Khetam Hussein – Analysis and interpretation of data, Drafting the article, Final approval of the article

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Spontaneous subcapsular renal hematoma in a patient being treated with dual antiplatelet therapy: A case report

Chrysovalantis Toutziaris, Kampantais Spyridon, Laskaridis Leonidas, Michail Koptsis, Filaretos Sardaridis, Stavros Ioannidis

ABSTRACT

Introduction: Spontaneous subcapsular renal hematoma is a relatively uncommon entity. After diagnosis, the determination of its cause is critical for the appropriate management. Case Report: In this report, we describe a case of spontaneous subcapsular renal hematoma in a patient without history of trauma. The patient was under double daily antiplatelet medication because of coronary heart disease. Conclusion: Diagnosis of the condition is suggested by ultrasound scan and confirmed by computed tomography (CT) scan. Literature suggests that the majority of these cases occur in association with renal tumors, and radical nephrectomy is recommended. When an underlying cause cannot be found, conservative treatment is proposed. However, the assessment must be completed with long-term, close surveillance, due to the risk of an undiagnosed neoplastic lesion. In our case, according to the negative imaging, the double antiplatelet therapy may be the only predisposing factor for the hemorrhage. It must be emphasized that discontinuation of medication and close follow-up can save the kidney.

Keywords: Spontaneous subcapsular renal hematoma, Antiplatelet therapy

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INTRODUCTION

Spontaneous subcapsular renal hematoma is defined as the hemorrhage of the renal parenchyma that is confined in the subcapsular space [1]. It is not a common entity in clinical practice. Till date, the rupture of a renal tumor was considered to be its most common cause and the recommended treatment was radical nephrectomy for non-fatty lesions [2, 3]. We report our experience with a patient with no other obvious cause for the hemorrhage except from his treatment with double antiplatelet medication. Our patient was treated conservatively with good outcome.

CASE REPORT

A 64-year-old male presented to the emergency department of our hospital with complaints of acute pain located at his left upper lateral midback, subcostally, mimicking renal colic pain. The patient did not give any history of trauma. However, he was under double daily treatment with antiplatelet medication, consisting of clopidogrel 75 mg and acetylsalicylic acid 100 mg. He was taking this medication because he had recently suffered from an acute myocardial infarction and had been subjected to coronary angioplasty a month ago.

During physical examination, his temperature was normal and there was mild tenderness situated at the

left costovertebral angle. Routine urine examination revealed microscopic hematuria. The laboratory examinations were normal, with initial hematocrit (Ht) 43%. The platelet count was 391x10^3/μL. No further platelet aggregation studies were performed. Kidney-ureter- bladder (KUB) radiography did not reveal any radio-opaque stone, while the initial ultrasound imaging revealed a subechogenic collection of fluid at the posterior renal surface. For this reason an abdominal computed tomography (CT) scan was immediately performed, showing extensive subcapsular hematoma of the left kidney with partial diffusion of blood in the perineal space (Figure 1). No other renal lesion was identified.

Because of the patient's haemodynamic stability, it was decided that conservative treatment was preferable with strict bed rest, rehydration, analgesic and antibiotic therapy (cefuroxime 750 mg three times daily I.V, paracetamol 1000 mg three times daily I.V). After consultation with cardiologists, the antiplatelet medication was replaced with low molecular weight heparin (tinzaparin sodium 14000 IU once daily S.C). The patient's symptoms receded within three days, while a gradual decrease of Ht up to 33% was noted. The patient was undergoing daily comparative ultrasound and the abdominal CT scan that was repeated seven days after the original CT scan, revealed constant stable findings without extension or worsening of the hematoma. The ultrasound that was performed after one month, showed a partial recession of the hematoma (Figure 2), while the abdominal CT scan six months later, revealed its almost total dissolution (Figure 3).

DISCUSSION

Spontaneous subcapsular renal hematoma was initially described by Bonet in 1679 [4]. Most common reported cases of spontaneous subcapsular renal hematoma in the absence of trauma include tumors (61.5%), vascular diseases, infections, peritoneal dialysis and post extracorporeal shock wave lithotripsy (ESWL). In 6.7% of cases the etiology cannot be found [5]. The incidence of tumors may be higher due to the fact that small, less than 2 cm in size tumors are undetectable by imaging techniques, including CT scan or angiography [6]. The most common associated renal tumor is angiomyolipoma (54%) followed by renal cell carcinoma (21%) [7].

Despite the fact that spontaneous subcapsular renal hematoma can present with Lenk's triad consisting of flank pain, tenderness and symptoms of internal bleeding, many other symptoms have been described in literature [2].

Ultrasound is very valuable in rapid identification of a renal hematoma. However, the findings should be confirmed with CT scan which also gives more

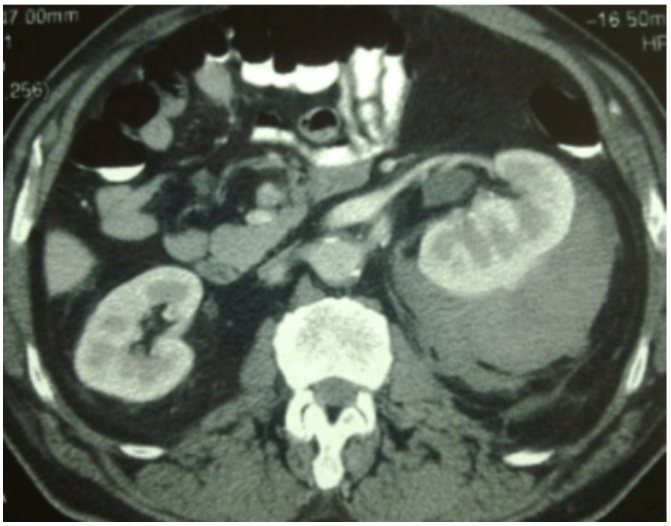

Figure 1: Initial contrast CT scan. A large subcapsular hematoma is observed in the left kidney.

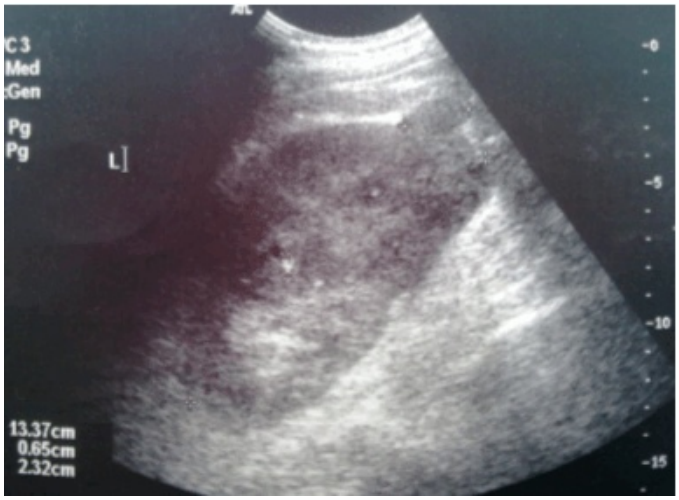

Figure 2: Ultrasound revealing partial recession of the hematoma in the first month after the hemorrhage.

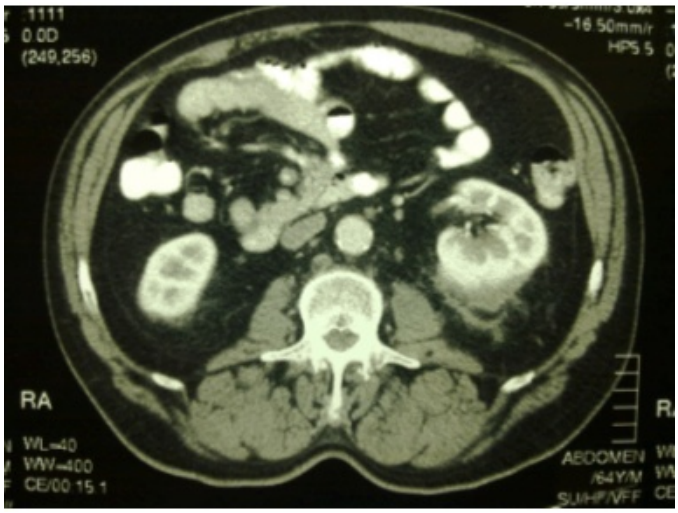

Figure 3: Contrast enhanced CT scan six months later showing almost total dissolution of the hematoma.

information regarding the cause of hematoma [4]. In our case there was no obvious responsible etiologic factor according to this imaging and the only cause that can be regarded is the dual antiplatelet medication of the patient. Captitanini et al. described a similar case of spontaneous subcapsular hematoma of the left kidney in a 67-year-old male. His therapy included acetylsalicylic acid (100 mg/day). A thoroughly investigation of his coagulation pattern was carried out revealing a platelet defect [8].

There are two possible treatment options regarding the optimal management of a spontaneous subcapsular renal hematoma. The first recommends radical nephrectomy because of the high incidence of small renal tumors especially in hemodynamically unstable patients. On the other hand, like in our case, conservative treatment is proposed to patients when an underlying pathology can be ruled out [2].

CONCLUSION

The cause of spontaneous subcapsular renal hematoma might not be evident in some cases. According to our case, the double antiplatelet may be the only predisposing factor and this rare complication may be attributed to this medication. Discontinuation of medication and close follow-up can save the kidney.

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Author Contributions

Chrysovalantis Toutziaris – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Kampantais Spyridon – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Laskaridis Leonidas – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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Stavros Ioannidis – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

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Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Wegener's granulomatosis with very unusual presentation

Mansur Somaily, Abdurhman S Al Arfaj

ABSTRACT

Introduction: Wegener's granulomatosis (WG) involves mainly upper and lower respiratory tract and the kidneys. Cardiac involvement is not common. Case Report: This is a report of a 34-year-old, non-smoker male with very unusual presentation of (WG) namely a combination of large pericardial effusion, cardiac tamponade (without uremia) and conduction defects, all of which responded very well to immunosuppressive therapy without the need for surgical intervention. Conclusion: Cardiac complication of WG may start early and early diagnosis and management improves the outcome and decrease, overall mortality.

Keywords: Wegener's granulomatosis, Cardiac involvement, Pericardial effusion, Cardiac tamponade

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INTRODUCTION

Wegener's granulomatosis is one of the most common forms of systemic vasculitis, with a reported annual incidence of 10 cases per million. The disease involves small and medium-sized blood vessels, and mainly affects the upper and lower respiratory tract and kidneys [1].

Cardiac involvement is reported to occur in 6–44% cases of Wegener's granulomatosis [2–5]. Pericarditis is the most common cardiac manifestation accounting for about 50% of cardiac diseases in Wegener's granulomatosis, which is asymptomatic in most of the cases, or may be manifested by chest pain and dyspnea [2–5].

Pericarditis and coronary vasculitis are the most frequent findings but myocarditis, endocarditis, valvulitis, and conduction system defects are also described [2–7]. Pericardial effusion reported in Wegener's granulomatosis can be due to the disease itself or due to uremia in cases of renal failure [7, 8]. We are reporting here a very unusual presentation of Wegener's granulomatosis which is a large pericardial effusion (without evidence of uremia), and cardiac tamponade combined with a conduction defect. This combined presentation has not been reported previously.

CASE REPORT

Our patient is a 34-year-old non-smoker male, who was admitted to the emergency room with history of chest pain and cough for five days, and fever for one month. His cough was mostly productive of whitish sputum but he had five attacks of hemoptysis of fresh blood of moderate amount (about 10 mL per day). He

also had one month history of arthralgia and daily epistaxis. He had no history of weight loss, nor contact with any patient of tuberculosis.

When he presented, he had a temperature of 37.8°C, but was hemodynamically stable. Examination of the nose revealed a nasal ulcer and tenderness over the left maxillary sinus. Skin, chest, cardiovascular and musculoskeletal examination were unremarkable. The initial differential diagnosis included: pulmonary tuberculosis, pneumonia, acute bronchitis and WG. His initial laboratory investigations showed WBC count $11x10^3$/L, Hb 13.6 g/dL, platelets $560x10^3$/µL, urea 5.3 mmol/L and creatinine 61 µmol/L. His erythrocyte sedimentation rate (ESR) was 120 mm/1st hr, aspartate aminotransferase (AGOT) 50 U/L, alanine aminotransferase (SGPT) 101 U/L, alkaline phosphatase 198 U/L and gamma glutamyl transferase (GGT) 232 U/L. His bilirubin, PT and APTT were normal. Cytoplasmic antineutrophil cytoplasmic antibody (cANCA) was 1:160 (normal range <1:40), and anti-proteinase 3 (anti-PR3) was 97 U/mL. Anti-myeloperoxidase (anti-MPO) and ANA were negative. Urine analysis showed hematuria with RBCs of 80/hpf, no WBCs and dipstick examination revealed 1+ positive protein. The 24-hour urine collection for protein was 0.5 g/day. Tuberculin test (5 units) was negative and sputum culture for acid fast bacilli was also negative. Chest X-ray was reported to be normal (Figure 1), but computed tomography (CT) scan of the lungs showed multiple nodules in both upper lobes, one of which was cavitating (Figure 2). CT scan of sinuses showed thickening of lining of left maxillary sinus. A nasal biopsy was obtained and showed necrotizing granulomatosis, consistent with diagnosis of WG. After two weeks of work-up the patient was diagnosed as WG based on clinical picture, cANCA positivity and tissue biopsy result. The patient was started on oral prednisolone 1 mg/kg/day and given one gram of intravenous cyclophosphamide. On the next day, the patient developed severe chest pain, dyspnea and hemoptysis. On examination, he was found to have high jugular venous pulse (JVP) with positive Kussmal's sign, pulsus paradoxus and distant heart sounds. The chest X-ray at that time showed-flask shaped heart (Figure 3). Echocardiography and CT scan of chest showed large pericardial effusion (Figure 4). The large pericardial effusion tamponade was thought to be most likely due to cardiac involvement by WG and as such, he was switched from oral prednisolone to one gram of intravenous pulse methylprednisolone. The next day patient developed light-headedness and dizziness, and was found to have bradycardia, with heart rate dropping down to 32 beats per minute (Figure 5). He was assessed by the cardiology team who advised continuous chronotropic monitoring and bed rest. He was continued on intravenous methylprednisolone one gram daily for three days which was reduced to 30 mg three times daily for two weeks and then switched to oral prednislone which was tapered down to zero over the following six months. The heart rate returned to normal after 12 days.

Two weeks after the first dose of cyclophosphamide, the patient was asymptomatic, with normal heart rate and complete resolution of pericardial effusion on echocardiography. He was discharged home on prednisolone, to be admitted for further five pulses of intravenous cyclophosphamide. He remained free of symptoms at follow-up.

DISCUSSION

Echocardiographic abnormalities were found in 80% patients with Wegener's granulomatosis but lesions were considered as related to Wegener's granulomatosis

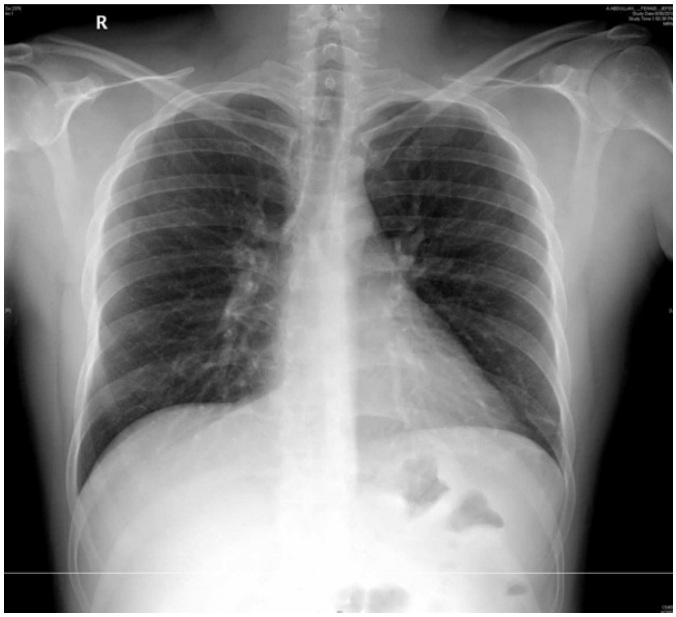

Figure 1: Chest X-ray was reported to be normal.

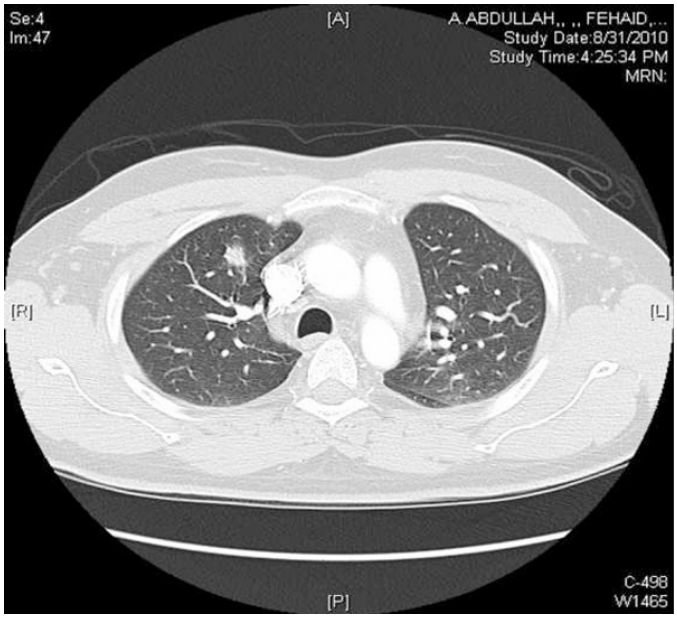

Figure 2: Computed tomography (CT) scan of the lungs showing multiple nodules in both upper lobes, one of which was cavitating.

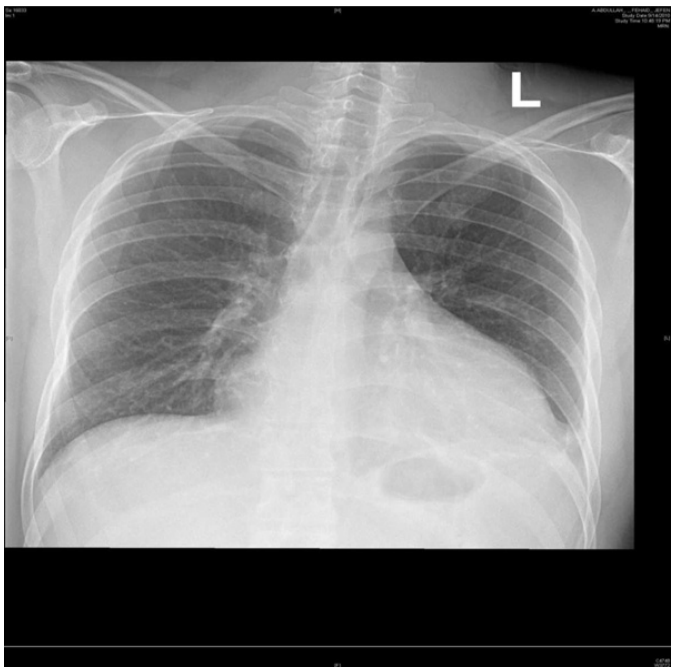

Figure 3: Chest X ray showing flask shape heart.

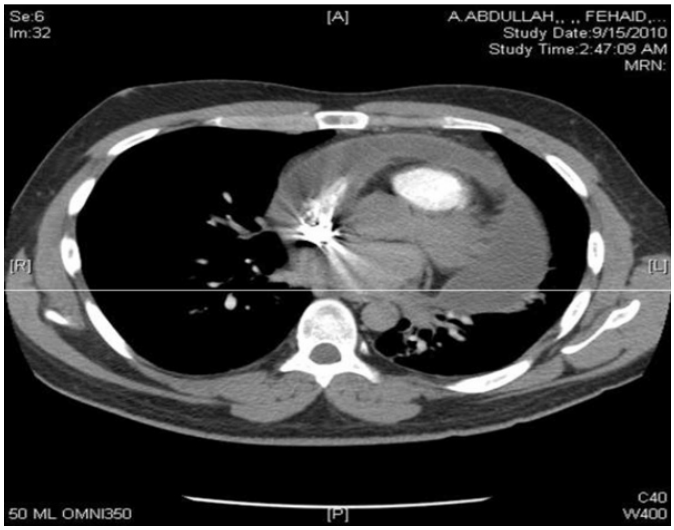

Figure 4: CT scan chest, showing large pericardial effusion.

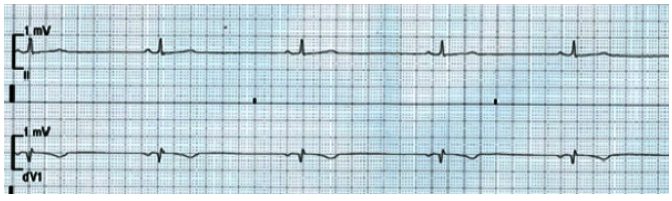

Figure 5: ECG demonstrating bradycardia, with heart rate of 32 beats per minute.

only in one-third of the patients [9]. Pericardial involvement represents almost 50% of cardiac involvement of Wegener's granulomatosis [5]. These are usually asymptomatic and carry a good prognosis. However some cases may lead to considerable morbidity and motality. The overall mortality rate of Wegener's granulomatosis with cardiac involvement has been reported to be between 15–45% [9].

Most of Wegener's granulomatosis patients reported previously with pericardial tamponade had hemodialysis dependant renal failure resulting in constrictive pericarditis [2–5, 7, 8]. However, massive pericardial effusion without constrictive pericarditis or uremia were reported in two cases [3, 4] .

The peculiar occurrence in our patient of large pericardial effusion and tamponade combined with conduction defect in the absence of any renal affection, has not been reported previously in any Wegener's granulomatosis patients. Cardiac tamponade in patient with Wegener's granulomatosis and renal failure are described before [2, 7]. Pericardial effusion per se is not so rare as a cardiac manifestation of Wegener's patients [10]. In a series of eleven Wegener's granulomatosis patients, who had transthoracic echocardiography (TTE) and cardiac magnetic resonance (CMR), pericardial effusion was observed in five patients, while localized pericardial thickening was seen in six patients [11]. Similarly, mild pericardial effusions were described in five out of nine Wegener's patients [5]. However, the degree of inflammation did not reach the stage of excessive effusion leading to tamponade. Conduction defects are also described in cases of Wegener's granulomatosis [12, 13]. However, the combination of both cardiac tamponade and conduction defects in the absence of uremia have not been described previously. We believe that the extreme intensity of the inflammatory procees of Wegener's granulomatosis in our patient resulted in a such an unusual combination of rare manifestations. This inflammatory process may be severe and lead to permanent damage. This was reported by Ohkawa et al., in a 61-year-old female with Wegener's granulomatosis who and developed by recurrent episodes of ventricular tachycardia developing bradycardia. Her autopsy revealed generalized necrotizing angitis and severe granulomatous inflammatory foci affecting the common bundle of His and right bundle branch in addition to the myocardium [14].

We do not believe that the occurrence of such combined presentation was due to a delay in the diagnosis or instituting treatment, as it took only six weeks to do so. On the contrary, it could be considered early diagnosis and treatment, as one of the largest series on Wegener's granulomatosis reported that the median and mean period from disease onset to diagnosis of Wegener's granulomatosis were 4.7 and 15 months, respectively [15]. That report found only ten cases of pericarditis among 158 cases of Wegener's granulomatosis [6%] and only one patient had large effusion requiring surgical intervention [15]. Some authors recommend echocardiographic screening examination for all patients with active Wegener's granulomatosis [9].

It is also worth noting that, the cardiac involvement in our patient responded very well to

immunosuppressive therapy alone with complete resolution of pericardial effusion and return of heart rate to normal and without the need for surgical or invasive procedures.

CONCLUSION

Our case serves as an example that cardiac complications of Wegener's granulomatosis may start early and emphasizes the need for early diagnosis of cardiac involvement and their management, so that patient outcome can be improved and overall mortality decreased.

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Author Contributions

Mansur Somaily – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Abdurhman S Al Arfaj – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Rural medicine in a developing economy: Does the presence of orthodox practice guarantee safe patient care?

Ikpeme A Ikpeme, Anthonia A Ikpeme, Emmanuel Efa, Elijah Udoh

ABSTRACT

Introduction: Medical practice in the developing world presents challenges which are more marked in the rural communities. A low doctor–patient ratio combined with a firm belief in traditional and unorthodox practices challenges the role of orthodox medicine in these communities. Despite the documented complications of unorthodox practices, injudicious orthodox care plays a significant role in the development of preventable complications and potential poor outcomes in the healthcare of rural dwellers in resource challenged societies. This article presents a case of severe preventable complications following injudicious orthodox care in a rural African region. Case Report: An 80-year-old rural dweller presented with a three-week history of lower urinary obstructive symptoms complicated by sepsis, hydronephrosis and renal parenchymal damage. He had spent three weeks in an orthodox medical practice in a rural community. Urinary catheterization produced 1700 mL of frank pus. He was managed by warm lavage, broad spectrum antibiotics and a referral to urological care. He made a full recovery from the complications and was offered elective prostatectomy. Conclusion: This case report highlights the potential for preventable complications in orthodox medical care in the rural areas of the developing world. Whereas orthodox practitioners are quick to highlight the complications that attend unorthodox medical practices, there is a need for appropriate audit and continuing medical education of orthodox rural practitioners.

Keywords: Rural medicine, Developing economy, Safe patient care

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INTRODUCTION

Medical practice in many developing countries presents unique challenges. These challenges are even more marked in the rural communities of the developing countries. With a doctor–patient ratio below the standard by World Health Organization (WHO), recommend, the role of customs, taboos and age long belief systems coupled with attendant poverty ensures that orthodox medicine faces a stiff challenge from unorthodox and traditional practices in these communities [1]. While the WHO works towards institutionalizing traditional medicine in the health systems of the African region, many reports have documented the complications that follow unorthodox and traditional practices in many of the world's poor regions [2–5].

Orthodox medicine has evolved based on scientific advances. One of the challenges we face today is the availability of high quality medical and health care to everyone at reasonable cost and with easy access [6]. A worldwide shortage of rural family physicians has been identified as contributing to difficulties in providing appropriate medical care in the rural areas of both developed and developing countries [7, 8]. In the rural areas of West Africa, the picture of medical practice has not changed much over the last 30 years [9]. Medical colleges producing an increasing number of orthodox physicians and with some of them setting-up practices in the rural areas it should be expected that the standard of care available to rural dwellers would improve. Our report shows that this may not be the case.

Trained in rigorous and structured training programs, orthodox physicians should bring the advantages of their training to bear on the healthcare status of the communities they work in. This must include safe practices, a recognition of the finiteness of their skills and early referral [9].

Access to safe healthcare presents an acute problem in the rural communities of many African countries. Despite huge investments in healthcare, there still exists significant urban–rural inequalities in healthcare in these communities. The index case demonstrates that among the indices that influence the healthcare of rural dwellers in Nigeria, the skills and knowledge base of orthodox practitioners are major contributors to outcomes. There needs to be a conscientious effort on the part of medical schools to develop curriculum to address rural practices and for regulators to encourage and enforce continuing medical education in these communities. This index case shows that beyond the often over flogged arguments for illiteracy, poverty, late presentation and injudicious unorthodox interventions as the underlying factors for poor health outcomes in the rural African communities, orthodox physicians need to examine their roles and the audit of their practices and interventions as a significant underlying factor in fostering poor outcomes. This report presents a case of severe preventable complications following injudicious orthodox care in a rural African region, and draw attention to the need for orthodox practitioners to focus on their practices as an important determinant of potential poor clinical outcomes in the developing countries.

CASE REPORT

AO, An 80-year-old rural dweller was presented to our practice with a three-week history of complete inability to pass urine, lower abdominal swelling and a general feeling of ill-health. Past medical history revealed features of lower urinary tract obstruction with straining, poor stream (not improved by straining) and a feeling of incomplete voiding lasting for about two years. There was no history of hematuria. The patient had presented in a rural private practice where he had spent the preceding three weeks. After one failed attempt at catheterization, he was managed by repeated supra-pubic bladder aspirations for three weeks. He received no antibiotics and was only referred when he became obtunded.

Clinical examination showed a weak, elderly man, with low body temperature (35°C), a respiratory rate of 30/min and an obtunded sensorium. There was bilateral pitting pedal oedema, generalized lower abdominal tenderness and the bladder size was 24 weeks. Abdomino-pelvic and transrectal ultrasonographic examination showed uniformly enlarged prostate (prostatic volume 85 cc) with no calcifications or suspicious foci, an enlarged bladder containing an estimated 1000 mL of fluid with fluid-fluid level, bilateral hydronephrosis and grade 3 renal parenchymal disease (Figures 1 and 2).

An impression of mismanaged bladder outlet obstruction with sepsis and reno-pelvic complications was made. The patient was managed by urethral catheterization which yielded 1700 mL of frank pus (subsequently becoming blood stained) (Figure 3), warm bladder lavage with 2000 mL normal saline, continuous bladder drainage with an in-dwelling catheter and intravenous broad spectrum antibiotics. A sepsis work up and serum electrolytes, urea and creatinine were requested and the patient was referred to the urologists. He subsequently made a full recovery from the immediate complications and was scheduled for elective prostatectomy.

DISCUSSION

Access to healthcare is a major problem in rural communities [10]. The World Bank group argues that despite major investments in health in Africa, populations in the poorer rural communities in Sub-Saharan Africa lose out in health service coverage [11]. One of the identified denominators of this position is the shortage of appropriate healthworkers in the rural

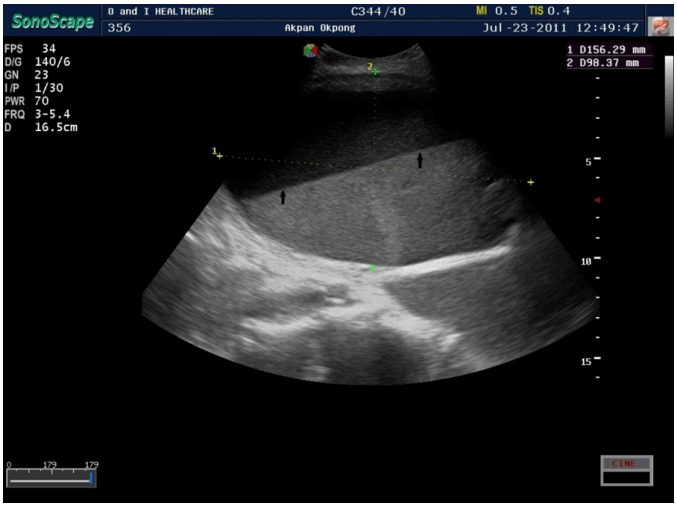

Figure 1: Ultrasound scan showed distended urinary bladder with fluid-fluid level (arrows).

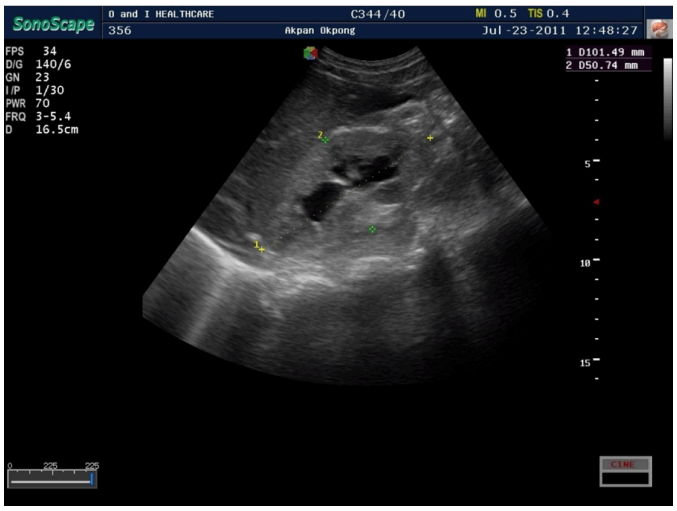

Figure 2: Ultrasound scan showed kidney with hydronephrosis and Grade 3 paranchymal disease.

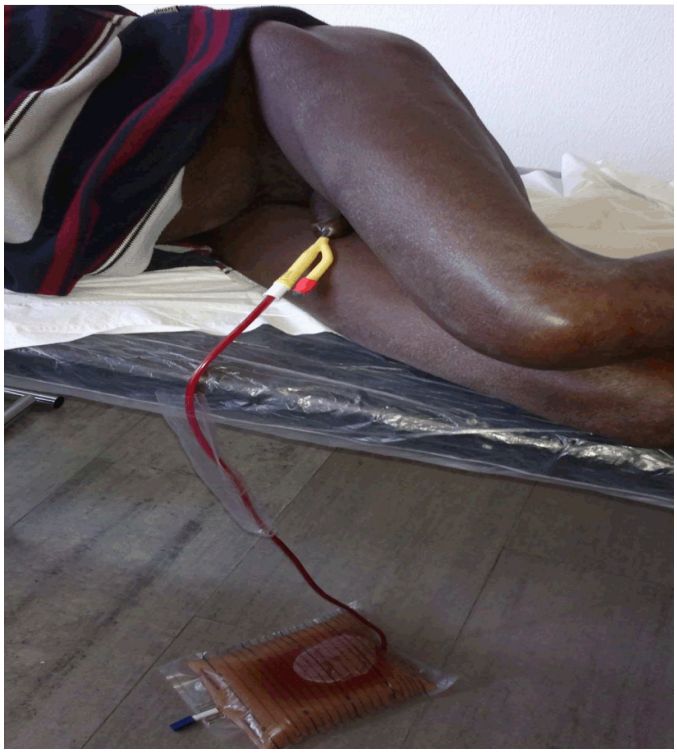

Figure 3: Catheter *in-situ* draining frank pus.

communities leading to urban–rural inequalities of healthcare [11, 12].

Access to an orthodox healthcare practitioner may not be the only challenge that rural dwellers face. The quality of healthcare available to rural dwellers is a function of many indices including the skills and knowledge base of the practitioners. The index case aptly demonstrates these issues. Whereas there are arguments for medical schools to help in the recruitment, training and retention of rural

Figure 4: Urine bag containing 1,700 mL of frank pus.

practitioners, [13–16] emphasis has to be laid on the quality of training available in medical colleges in the developing world. Graduating clinicians must be taught repeatedly that prompt and appropriate referral is a critical component of safe healthcare practices.

Like most other developing countries, the major proportion of Nigerians live in the rural areas. Patients also have a strong positive perception of traditional health practices but will accept orthodox care [17, 18]. While orthodox practitioners have documented the complications that follow traditional medical practices [1, 3, 17], orthodox medicine can only win the hearts of the majority of rural dwellers if consistent and proven safe outcomes are achieved. Medical schools, therefore, need to target the development of rural practices in their curricular [12–14, 19]. There is available evidence that medical schools can maintain competitive admission criteria while attracting and graduating students who will likely enter rural practice [19].

Continuing medical education aims to foster lifelong learning in a physician. This ultimately translates to the maintenance and increase in knowledge, skillbase and professional competence of physicians. This is well established in the developed world and only becoming implemented in Nigeria. Besides targeting the development of rural practice curricular, continuing medical education when enforced will help in the reduction of avoidable complications in orthodox care and argurably improve the quality of healthcare available to rural dwellers in the developing world.

CONCLUSION

Our report highlights the potential for preventable complications in orthodox medical care in the rural areas of the developing world. While orthodox practitioners highlight the complications that follow traditional medical and other unorthodox interventions, there is the need to encourage continuing medical education, early and appropriate referral and regular audit of orthodox medical practices in the developing countries. The inequalities in urban–rural healthcare should be addressed in ways that ensure consistently safe outcomes for rural dwellers.

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Author Contributions

Ikpeme A Ikpeme – Substantial contributions to conception and design, Acquisition of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published
Anthonia A Ikpeme – Substantial contributions to conception and design, analysis and interpretation of data, Drafting the article, Final approval of the version to be published
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Elijah Udoh – Substantial contributions to conception and design, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published

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The corresponding author is the guarantor of submission.

Conflict of Interest

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CASE REPORT OPEN ACCESS

An incidental mass in the inferior vena cava discovered on echocardiogram

Timothy R Larsen, Kate Essad, Sachin Kumar Amruthlal Jain, Mark Lebeis, Shukri David

ABSTRACT

Introduction: The differential diagnosis of an inferior vena cava (IVC) mass in an asymptomatic patient is broad and includes both thrombus and neoplastic etiologies. Once identified, new IVC masses should be further evaluated for possible malignancy. Up to 10% patients with renal cell carcinoma have direct extension of the tumor into the IVC, with 1% extending to the level of the right atrium. Case Report: A 73-year-old woman was referred for an elective two-dimensional echocardiogram after her cardiologist noted a new murmur. The patient was asymptomatic at that time. The echocardiogram revealed a mass in the IVC, two centimeters from the right atrium. Further work up revealed a large renal cell carcinoma. Conclusion: All imaging results should be carefully reviewed for incidental evidence of previously undiagnosed disease. Malignant renal cell carcinoma is often first identified as an incidental mass on abdominal CT scan or ultrasound scan, It is exceedingly rare for a malignant renal cell carcinoma to initially present as an incidental mass on echocardiogram.

Keywords: Inferior vena cava (IVC), Incidental IVC mass, Renal cell carcinoma, IVC tumor extension

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INTRODUCTION

Imaging studies frequently provide information above and beyond what is expected at the time of requisition. Unexpected diseases may be discovered prior to the development of symptoms and thus provide the opportunity for early diagnosis and intervention. Early treatment can often prevent or limit complications, including major morbidity and death. Koeneman et al. reported the prevalence of malignant renal tumors to be 0.2% in adults aged 50–79 years [1]. In many cases these tumors are discovered incidentally on imaging, usually by abdominal computed tomography (CT) scan or ultrasound scan [2]. The identification of a renal malignancy on routine echocardiography is exceedingly rare. Here we present a case where a large malignant neoplasm originating from

the right kidney was first identified as an incidental mass in the inferior vena cava on echocardiogram.

CASE REPORT

A 73-year-old female met with her cardiologist for a routine check-up. She was in her usual state of health; she denied dyspnea, chest pain, palpitations, syncope pre-syncope, shortness of breath, dysuria, and abdominal pain. All other review of systems was unremarkable. A new murmur was noted on physical examination, therefore a two-dimensional resting echocardiogram was done which revealed a mass in the inferior vena cava (IVC), two centimetres below the right atrium (Figure 1). The patient was admitted to the hospital for further evaluation. On admission her vital signs were: temperature - 36.4°C, respiratory rate - 20/min, pulse - 68/min, blood pressure - 134/69 mmHg, and BMI 40 Kg/m². Cardiac examination revealed a grade 2/6 crescendo-decrescendo murmur heard at the base that did not radiate, there were no gallops and point of maximal impulse was not displaced. The remainder of the physical exam was unremarkable. Laboratory analysis revealed hemoglobin of 10.8 g/dL and glomerular filtration rate of 42 mL/min. Rest of the laboratory investigations of CBC, electrolyte panel, urinalysis, thyroid function, and coagulation studies were within normal range. A CT scan of the chest and abdomen revealed a 10x13x15 cm mass arising from the anterior right kidney with direct extension into the suprarenal IVC (Figure 2). There was no evidence of distant metastasis. Pulmonary emboli (PE) were also noted in the right main pulmonary artery and right middle arteries. Intravenous heparin therapy was initiated. After one month of anticoagulation, repeat CT scan demonstrated no interval change of the mass. Three months after presentation, the patient underwent right nephrectomy with excision of a 12.5x11.5x10.5 cm mass which was confirmed to be renal cell carcinoma (RCC) with extensive necrosis and hemorrhage. Vascular and cardiothoracic surgeons assisted in the resection of the 14.5 cm long, 1.2 cm diameter, polypoid portion that extended into the IVC and was adherent to the inner lining of the vein. The IVC was cross clamped, opened, and the mass was resected. The IVC was then flushed, the incision was closed, air removed, and the clamps released with restoration of blood flow. The patient is well and is on regular follow-up.

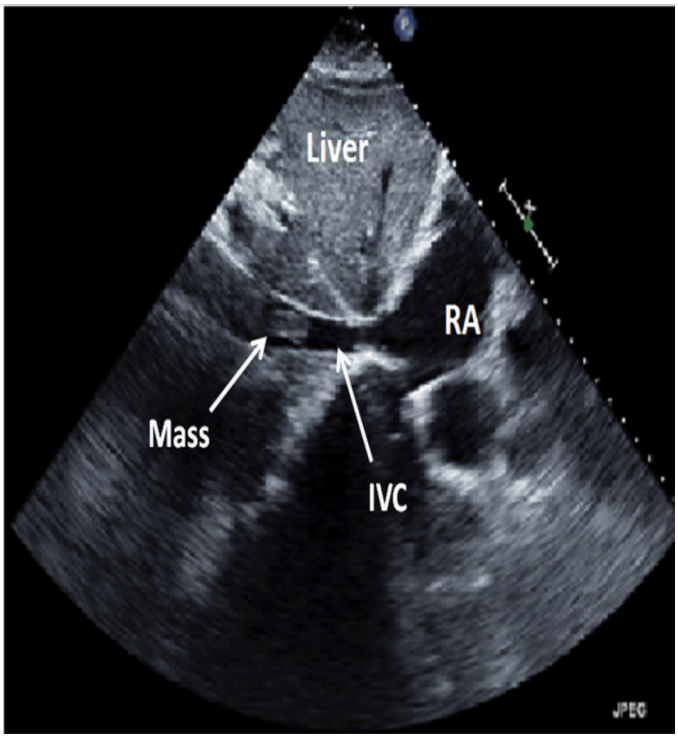

Figure 1: Two-dimensional echocardiogram showing a mass in the IVC two cm below the right atrium.

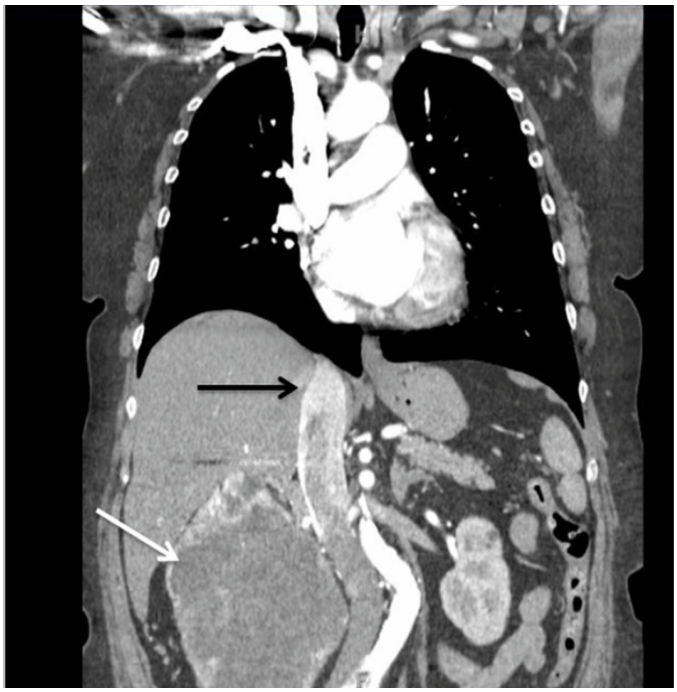

Figure 2: CT coronal image of the chest and abdomen in venous phase showing a 10x13x15 cm heterogeneous mass (white arrow) arising from the right kidney extending into the IVC (black arrow) (120 ml of Isovue-370 contrast, iv).

DISCUSSION

The differential diagnosis for an IVC mass includes: bland thrombus (most common), malignant thrombus, carcinoma (renal cell, hepatocellular, and adrenocortical), renal angiomyolipoma, pheochromocytoma, pseudolipoma, and primary intraluminal sarcoma [3]. A malignant etiology should be considered in all patients with a new IVC mass.

Renal cell carcinoma is associated with extension into the IVC in 4–10% of cases [4–6], with approximately 1% extending to the level of the right

atrium [5]. Up to 6% patients with direct extension into the IVC suffer pulmonary embolism which is a poor prognostic sign and is associated with a high rate of mortality [5].

Patients with RCC may present with microscopic hematuria, complaints of flank or abdominal pain and weight loss [4]. If a pulmonary embolism (PE) has occurred symptoms, typical of PE may present including dyspnea, pleuritic chest pain, hypotension, hemoptysis, and hypoxemia [7]. Dyspnea on exertion and syncope may present if the tumor extends to the level of the right atrium [8]. Serious complications of RCC with IVC and right atrium infiltration include massive PE, tricuspid valve obstruction, and Budd–Chiari syndrome [4]. This patient did not experience any of these associated symptoms.

Surgical intervention for RCC with IVC extension is the standard of care as complete surgical resection can potentially be curative [4]. Additionally, surgical intervention is required to prevent serious complications such as massive PE, tricuspid valve obstruction, and Budd–Chiari syndrome with resultant hepatic failure [4]. Operative technique is determined by the level of tumor extension following the Neves classification system (Table 1) [6]. The patient was Neves Level IV, thus warranting a multidisciplinary surgical approach including urological and cardiothoracic surgical evaluation. If the tumor had invaded the right atrium, cardiopulmonary bypass would have been recommended [6].

Table 1: Neves classification of renal tumor thrombus extension as adapted from Radak et al.

Level	Involvement
I	IVC at level of renal vein
II	Infrahepatic IVC
III	Retrohepatic IVC
IV	Supradiaphragmatic IVC or right atrium

Abbreviations: IVC - Inferior vena cava

CONCLUSION

This case is a unique example of an asymptomatic individual diagnosed with an advanced malignancy based on an echocardiographic abnormality and thus underscores the importance of careful and detailed review of all radiology investigations. The serendipitous discovery of this nephrogenic malignancy allowed for the initiation of potentially curative treatment before the development of serious complications including massive PE, tricuspid valve obstruction, and death.

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Author Contributions

Timothy R Larsen – Substantial contributions to Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Kate M Essad – Substantial contributions to Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Sachin Kumar Amruthal Jain – Substantial contributions to Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Mark Lebeis – Substantial contributions to Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Shukri David – Substantial contributions to Conception and design, Acquisition of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Lyme arthritis versus acute bacterial arthritis: A diagnostic dilemma

Dinushan Kaluarachchi, Magda Mendez, Shefali Khanna

ABSTRACT

Introduction: Lyme disease is a known cause of monoarticular arthritis in Lyme endemic areas. It must be differentiated from other types of acute monoarticular arthritis such as acute bacterial arthritis. Case Report: In this report, we discuss the case of a 9-year-old boy who presented to emergency room with left wrist monoarthritis of one day's duration. Initial laboratory evidence were suggestive of septic arthiritis and treatment was initiated for the presumed diagnosis of septic arthritis, but high index of suspicion lead to further investigations and diagnosis of Lyme arthritis. Conclusion: Diagnosis of Lyme arthritis may be difficult. Exposure in an endemic area and clinical findings may help distinguish it from acute bacterial arthritis.

Keywords: Lyme arthritis, Acute bacterial arthritis, Borrelia burgdoferi

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INTRODUCTION

Lyme disease is a common cause of monoarticular arthritis in Lyme endemic areas. Lyme arthritis is a late manifestation of Lyme disease, occurring in seven percent of patients with Lyme disease [1]. This form of arthritis must be differentiated from other types of acute monoarticular arthritis such as acute bacterial arthritis. This differentiation is important for treatment and prognosis. Failure to diagnose and treat Lyme arthritis can result in persistent joint symptoms and risk of developing additional disease manifestations. In this report, we discuss a patient who presented with wrist monoarthritis, who was initially treated as septic arthritis, but later turned out to be Lyme arthritis.

CASE REPORT

A 9-year-old previously healthy, African-American boy presented to emergency room (ER) with history of one day's swelling and pain of the left wrist joint. Movements of the affected joint were restricted due to pain. There was no history of fever, rashes or other joint involvement. Patient denied trauma or insect bites. He was treated in the ER one month back for left knee arthralgia with effusion attributed to a twisting injury. The symptoms were resolved following treatment with ibuprofen. Patient lived in Bronx, NY, but had traveled upstate New York on several occasions, once for a summer camp about six months prior to the presentation.

Swelling and tenderness over left wrist joint was noted on examination. Wrist X-ray revealed no fractures or dislocation. Complete blood count showed WBC count of $8.7x10^3/mm^3$ with 57% neutrophils and 40% lymphocytes. The ESR was 40 mm/1st hr. Joint aspiration was performed in the ER. Five milliliters of purulent fluid was drained. Synovial fluid analysis was significant for WBC count of $2.53x10^5/mm^3$ with 93% neutrophils. Blood and synovial fluid cultures were sent. Septic arthritis was diagnosed based on the clinical presentation and synovial fluid analysis results. Patient was started on IV ceftriaxone and vancomycin. The next day the patient underwent left wrist irrigation and debridement. Cultures of the blood and synovial fluid were subsequently reported negative.

The clinical diagnosis of septic arthritis was in doubt as the patient did not have fever, significant pain, leukocytosis or significantly high ESR. These findings and the history of left knee arthralgia with effusion one month prior directed to further investigations. Patient underwent further investigations to rule out other possible etiologies.

Serologic tests for *Borrelia burgdoferi* revealed presence of 9/10 *B. burgdoferi* specific IgG bands and 3/3 *B. burgdoferi* specific IgM bands on western blot. The diagnosis of Lyme arthritis was made and patient was discharged on oral amoxicillin to complete 28 days course of the antibiotic. At one week and one month of follow-up after discharge, the wrist pain and swelling had resolved and normal range of movements of the left wrist was observed.

DISCUSSION

Lyme disease is the most common tick borne disease in the US and Europe. *Borrelia burgdoferi* is the sole cause of the disease in the United States. Lyme arthritis is a late manifestation of Lyme disease, occurring in seven percent of patients with Lyme disease [1]. The arthritis is usually monoarticular or oligoarticular and affects the large joints, particularly the knee joint which is involved in more than 90% cases [2, 3]. There is a wide spectrum in the acuity of presentation of Lyme arthritis and in some instances it may be confused with acute bacterial arthritis [3, 4].

Clinical features which may help to distinguish Lyme arthritis from acute bacterial arthritis include [3, 4],

- In Lyme arthritis, the joint pain is less intense than that associated with bacterial arthritis.
- Most children with Lyme arthritis of the knee joint can walk without difficulty despite some limitation of range of motion.
- Associated fever and erythema of involved joint is less common in Lyme arthritis.
- In Lyme arthritis, synovial fluid WBC count usually ranges from $2–6x10^4/mm^3$ (but can exceed $1x10^5/mm^3$), whereas in acute bacterial arthritis, the synovial fluid WBC count is usually more than $1x10^5/mm^3$.
- If untreated, Lyme arthritis usually lasts for several weeks before resolving, only to recur often in a different joint [5]. Acute bacterial arthritis does not improve without treatment.

Serological responsiveness to *B. burgdoferi* is the primary laboratory test for diagnosing Lyme arthritis. Criteria for western blot immunoglobulin requires the presence of at least five out of ten specific bands [6]. For Lyme arthritis in the absence of neurologic disease, the recommended initial regimen is oral doxycyline or oral amoxicillin for 28 days [2]. The Infectious Diseases Society of America has suggested cefuroxime as an alternative agent in those with contraindications to doxycycline or amoxicillin.

CONCLUSION

The diagnosis of Lyme arthritis may be difficult. Exposure in an endemic area and clinical findings may help in distinguishing it from acute bacterial arthritis.

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Author Contributions

Dinushan Kaluarachchi – Substantial contributions to analysis and interpretation of data, Clinical patient care, Drafting the article, Final approval of the version to be published
Magda Mendez – Substantial contributions to analysis and interpretation of data, Clinical patient care, Drafting the article, Final approval of the version to be published
Shefali Khanna – Substantial contributions to analysis and interpretation of data, Clinical patient care, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Unusual case of pancreatic ascites and pancreatic pleural effusion following endoscopic retrograde cholangiopancreatography

Rafael Alba Yunen, King Soon Goh, Ugoagha Chimbo-Osuagwu, Sulaiman Azeez

ABSTRACT

Introduction: Pancreatic fistula is the most common complication of pancreatic injury in the setting of blunt trauma and chronic alcoholic pancreatitis. Internal pancreatic fistulas (IPF) are most commonly cause by disruption of the pancreatic duct due to pancreatitis, and leakage from a pancreatic pseudocyst. We present a case of post-endoscopic retrograde cholangiopancreatography (ERCP) pancreatic leak complicated by pancreatic pleural effusions and pancreatic ascites. Case Report: An 18-year-old Hispanic female was admitted with persistent right upper quadrant and epigastric pain, abdominal distention, constipation, leukocytosis and elevated pancreatic enzymes, with history of recent laporoscopic cholecystectomy due to cholelithiasis three months ago. She was diagnosed with choledocholithiasis, underwent successful ERCP, and was discharged home. After 24 hours of discharge, she developed symptoms of systemic inflammatory response syndrome and constipation. Computed tomography (CT) scan of abdomen and paracentesis revealed left sided ascites and plural effusion with high amylase content and no infection. She was diagnosed as suffering from pancreatic ascites and pancreatic pleural effusion following endoscopic retrograde cholangiopancreatography, secondary to internal pancreatic fistula. She received octreotide and a conservative approach to her condition, and had a good response to therapy. Conclusion: In our patient diagnosis was made by finding elevated amylase and protein content in the ascitic and pleural fluids plus CT scan revealed effusion and ascites and managed conservatively. The use of octreotide in such cases is established, and has been successful in our experience.

Keywords: ERCP, Internal pancreatic fistula, Pancreatitis, Pancreatic ascites, Plueral effusion

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INTRODUCTION

Pancreatic fistula is the most common complication of pancreatic injury in the setting of blunt trauma and chronic alcoholic pancreatitis [1]. Internal pancreatic fistula are most commonly cause by disruption of the pancreatic duct due to pancreatitis, and to a less extent by leakage from a pancreatic pseudocyst. When the disruption is anterior, an internal pancreatic fistula

(IPF) develops into the peritoneal cavity causing pancreatic ascites. If the disruption is posterior and secretions track up into the mediastinum, pleura is penetrated and an IPF into one or both cavities is established causing pancreatic pleural effusions. Endoscopic retrograde cholangiopancreaticography (ERCP) is one of the procedures that explores the branches of the biliary tree, by means of injecting contrast material via the sphincter of Oddi which could potentially damage the wall of the biliary tract.

The ERCP has been the initial modality of intervention for retained stones in the common bile duct (CBD) in patients scheduled for laparoscopic cholecystectomy. Although the procedure is usually safe, procedure related complications do occur, the most serious of which are perforation, bleeding and pancreatitis. Necrotising pancreatitis, pseudocysts, pancreatogenic ascites and infection have been reported. Systemic complications leading to multi organ failure are the usual cause of death in cases of pancreatitis.

The diagnosis of pancreatic fistula is usually made in a typical setting of unexplained persistent elevation of amylase and protein content in the ascitic and pleural content and by using computed tomography (CT) scan and ERCP. Post-ERCP pancreatitis is usually mild and self-limiting. Treatment options are directed on decreasing the flow across the leak and increased resistance, which can be achieved non-surgically with the use of long acting somatostatin analogues for approximately 2–3 weeks to reduce the flow, with pancreatic end prosthesis or surgery. Although spontaneous closure of pancreatic ductal disruption has been reported, surgical treatment is accepted as the single most common intervention in major ductal injury but has considerable morbidity and mortality.

Here we present a case of post-ERCP pancreatic leak complicated by pancreatic pleural effusions and pancreatic ascites.

CASE REPORT

An 18-years-old Hispanic female was admitted to our institution with history of intermittent right upper quadrant pain, abdominal distention, nausea and vomiting consistent with biliary colic. These symptoms were occasionally triggered by ingestion of fatty foods. Given her initial presentation suggestive of classical gastritis, an esophagogastroduodenoscopy (EGD) was done with unremarkable results. Laboratory investigations showed no signs of cholestasis. Her symptoms were persistent which warranted further workup. There was evidence of cholelithiasis on CT scan of abdomen (Figure 1). Laparoscopic cholecystectomy with intra-operative cholangiogram was performed with findings consistent with an acutely inflamed gallbladder. She was discharged home two days after the procedure. However, she was admitted three months later with recurrent right upper quadrent (RUQ) pain, nausea and vomiting associated with food intake. Her liver function test (LFT) were abnormal. The common bile duct (CBD) was dilated with stones on liver ultrasonography, consistant with choledocholithiasis. She underwent successful ERCP and sphincterotomy with a stent placement by gastroenterologist with stone extraction on day-2 of admission. There were no acute changes in her condition and she was subsequently discharged home the next day on oral antibiotics, with improving clinical signs and liver function test (LFT).

After about 24 hours, she was re-admitted with persistent RUQ and epigastric pain, abdominal distention and constipation. She was found to have elevated pancreatic enzymes, leukocytosis and dilated loops of bowel without air levels on abdmominal X-ray consistent with ileus (Figure 2). At the same time, there was evidence of a small left pleural effusion and ascites on CT scan of abdomen (Figure 3). With the assistance of interventional radiology, a diagnostic paracentasis was performed. Her clinical condition continued to deteriorate with tachycardia, persistent leukocytosis, worsening LFTs and pancreatic enzymes, and with serum amylase peaking at 2,097 U/L. She was transferred to MICU for close monitoring; and was treated for opiod induced ileus and constipation, with non-pathologically raised pancreatic enzyme post-ERCP. Fluid analysis from paracentasis later showed markedly elevated amylase of 46,250 U/L, without significant bile (bilirubin 1 mg/dL; LDH 2,345 U/L) or pus. There was no indication of drain insertion as both pleural effusion and ascites were too small to be drained. Instead, she was managed conservatively with bowel rest and octreotide (100 μg subcutaneous) was started along with broad-spectrum antibiotic coverage with imipenem/cilastatin. Blood cultures were negative for bacterial growth. Her antibiotic therapy was stopped after four days of treatment. Enteral feeding via a tube in proximal jejunum was started in time without complication. Her clinical condition further improved with no fever and her abdomen became non-tender on palpation. CT scan of abdomen showed improvement with CBD reduced in size (Figure 4). The patient had normal stool formation. The opiod dose was tapered and normal oral feeding was re-started. A diagnosis of internal pancreatic fistula/pancreatic duct leak with pancreatic ascites and pleural effusion was made.

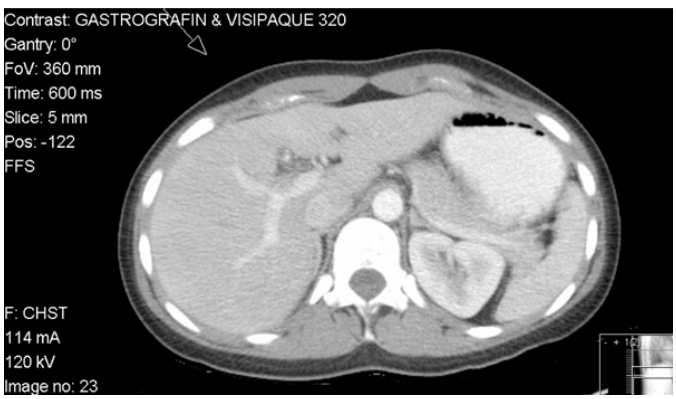

Figure 1: Computed tomography scan of abdomen and pelvis showed gallstones with sludge (gastrografin and vasipaque320, power 114 mA, 120 kV).

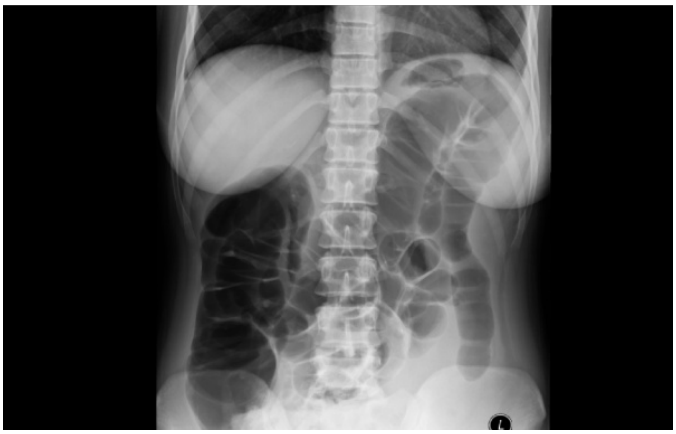

Figure 2: Abdominal X-ray showed distended loops of bowel suggestive of ileus.

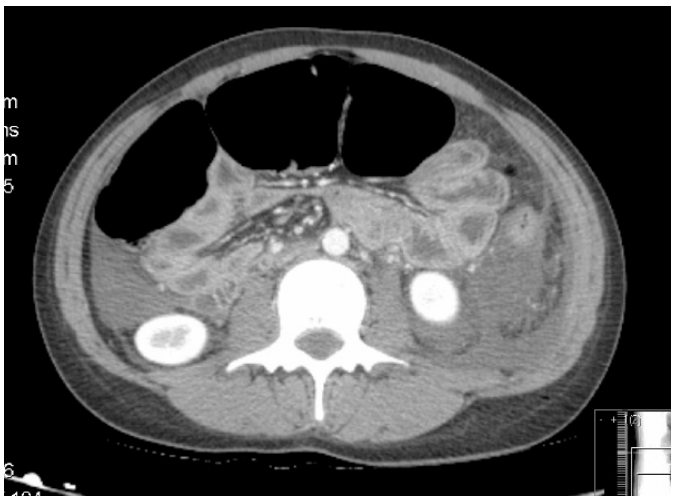

Figure 3: CT scan of abdomen and pelvis without contrast showed peritoneal, left retroperitoneal and pelvic ascites with likely ileus power 140 mA, 120 kV.

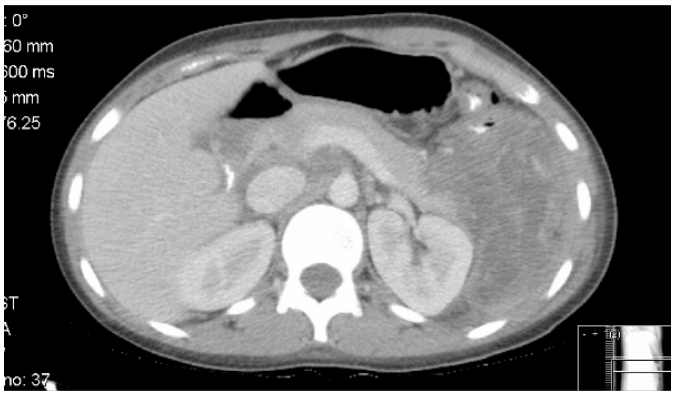

Figure 4: CT scan of abdomen and pelvis showed hepatic and common bile duct reduced to normal size gastrografin Isovue 300; power 123 mA, 120 kV.

DISCUSSION

This case report highlights an important issue of the potential risk of ERCP associated complication of pancreatic leak leading to internal pancreatic fistula (IPF). Ordinarily when a pancreatic duct disruption occurs, it is during an attack of acute pancreatitis and there is enough inflammatory reaction so that the disruption is walled off by the stomach, transverse colon, mesocolon and other surrounding tissues and a pseudocyst is formed. When a duct disruption occurs in the absence of acute pancreatitis, the duct disruption is not walled off, and an IPF results. The incidence of post-ERCP pancreatitis is reported to range from 1.3–6.7%. Post-ERCP pancreatitis is defined as abdominal pain for more than 24 hr after the procedure and levels of serum pancreatic enzymes three times above normal. Pancreatitis and high serum amylase usually occur after difficult procedures in which pancreatography was performed [2]. The elevation of serum amylase is not a specific sign of pancreatic injury. Small CBD and pre-cut papillotomy also significantly increases the risk of pancreatitis [2]. ERCP has a major complication rate of 1.38% for diagnostic and 5.4% for therapeutic purposes [3].

The diagnosis is made by routinely sending all pleural effusions and ascitic fluid for amylase and albumin determination. Amylase will be markedly elevated, not necessarily elevated in the serum, and in the absence of hypoalbuminemia, albumin will be over 3 g/dL in the ascitic or pleural fluid, which will be diagnostic. Pancreatic ascties diagnostic criteria include ascitic fluid amylase >2000 IU/L in >5 simultaneous values. Recent articles in literature suggest that CT scan has 90% sensitivity for detecting pancreatic ductal disruption [4]. Gougeon et al. [5] first reported the use of emergency ERCP in the diagnosis of pancreatic injury in 1976. ERCP has a sensitivity and specificity of 100% for pancreatic ductal injury [6]. ERCP provides not only a conclusive diagnosis but also an effective and safe non-operative treatment tool, in applications such as sphincterotomy, removal of CBD stones, lithotripsy, biliary drainage, and stricture dilation [7]. However, ERCP requires a stable patient and a skilled endoscopist, and has its own complications like pancreatitis, infection, duodenal injury and pancreatic duct disruption.

Despite a success rate of only 48% and a mortality of 16%, non-operative therapy should probably be attempted as the initial treatment in all patients with the IPF. Treatment of leak has focused on decreasing the flow and increasing resistance like a drain removal or fibrin glue. Most authorities feel that the pancreatic fistulas should be managed conservatively as the majority close within a month and operative management should be reserved for the failure of conservative management, peritonitis and associated duodenal injury. Some case series have shown pancreatic duct stent placement to be an effective therapy in resolving duct disruption. Transductal pancreatic stenting allows internal drainage of the pancreatic secretion and may re-establish duct continuity, although a proportion still requires percutaneous or endoscopic drainage. Endoscopic management with a stable patient and a skilled

endoscopist has gained increasing acceptance over the past decade. Endoscopic sealing of fistulas by endoscopic injection of the sealant have also been used.

Our patient underwent successful ERCP and sphincterotomy with a stent placement for protection of the pancreas, stone extraction and stent removal. Stent placement is known to decrease the incidence of ERCP directed pancreatitis. Post-ERCP our patient had elevated pancreatic enzymes typical of post-ERCP pancreatitis, however, CT scan showed septated complex collection in perisplenic and tail region of the pancreas. Septated collection represents walling at the point of pancreatic duct leak which does not occur with post-ERCP pancreatitis in which the entire pancreas is edematous. In case of a possible disruption at the level of the tail, it was most likely due to the guide wire insertion with distal perforation. Another important factor was constipation and dilated loops of bowel, 'ileus' which was likely multifactorial from the use of opiods and pancreatic ascitis. There has been reported cases of such similar nature [8].

Our patient's diagnosis was made by finding elevated amylase and protein content in the ascitic and pleural fluids plus CT scan revealing effusion and ascites and managed conservatively. The use of octreotide in this case was established, and has been successful in our experience. Nonetheless the latest literature concluded that octreotide does not aid in the resolution of the fistula. Options for treatment of a persistent chronic fistula include removal of the drain and injection of the fistula tract with fibrin glue or fistula tract-enteric anastomosis.

CONCLUSION

The long-term outlook for patients with pancreatic ascites and pancreatic pleural effusions is certainly much more favorable than that of entities with which they are usually confused with like cirrhotic ascites and lung cancer. Although a pancreatic leak is a very rare complication, it can become serious condition adding to morbidity and mortality.

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Author Contributions

Rafael Alba Yunen – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

King Soon Goh – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

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Sulaiman Azeez – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CASE REPORT OPEN ACCESS

Umblical hernia, hypertelorism, sensorineural deafness: Is it Donnai–Barrow syndrome?

Serap Tutgun Onrat, Yaşar Sivaci, Ersel Onrat, Tülay Köken

ABSTRACT

Introduction: Donnai–Barrow syndrome is a rare autosomal recessive disorder first described in 1993 and characterized by diaphragmatic hernia, hypertelorism, agenesis of the corpus callosum and deafness. Case Report: A 23-year-old female with clinical features similar to Turner patients were sent to our department by cardiology department. The main features were umblical hernia, hypertelorism (interpupillary distance 45 mm), and sensorineural deafness. Other findings included mid-face hypoplasia, a broad forehead, exotropia, frontal bossing and wide anterior fontanel, down slanting palpebral fissures, short nose with a broad tip, cubitis valgus and posterior rotated ears, webbed neck and short stature (height - 144 cm, weight - 47.5 kg). Magnetic resonance imaging (MRI) scan of the brain confirmed arachnoid cyst, cervical spinal stenosis (thought to be secondary to hydrocephalus) and absent corpus collosum. Autosomal recessive inheritance was suspected because patient's parents were also first cousins. Cytogenetic analysis demonstrated normal karyotype (46, XX). Conclusion: We describe a female patient who shares identical characters to the patients described by Donnai and Barrow. Although our patient has got a large number of malformations, her karyotype was normal, which makes this case extremely interesting. Such patient's have moderate levels of mental life which is consonant with the disease, and the patients can adapt to social life.

Keywords: Donnai–Barrow syndrome, Autosomal resessive inheritance, Normal karyotype, Umblical hernia, Hearing loss

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INTRODUCTION

Donnai and Barrow reported two sets of siblings and an unrelated individual with a new syndrome. [1] Although karyotype is normal, some typical clinical features of the Donnai–Barrow syndrome overlap the phenotype of the 9q microdeletion syndrome [2]. The phenotype associated are diaphragmatic hernia, exomphalos, absent corpus callosum, intestinal malrotation, myopia, sensorineural deafness, and particular face. Donnai–Barrow syndrome

Faciooculoacousticorenal syndrome (DB/FOAR) is a rare autosomal recessive disorder resulting from mutations in the LRP2 gene located on chromosome 2q31.1 [3]. Small deletions or insertions causing frameshifts, as well as conserved splice site, nonsense and missense mutations of low-density lipoprotein receptor related protein 2 (LRP2) gene in seven DBS/FOAR families were recently reported [4]. The 79 exon LRP2 gene mapping to human chromosome 2q31.1 encodes megalin, an endocytic transmembrane glycoprotein [5].

CASE REPORT

A 23-year-old female, with clinical features similar to Turner patients was referred to our department by the cardiology department. The echocardiographic examination was normal. The main features were umblical hernia (Figure 1), hypertelorism (interpupillary distance 45 mm) and sensorineural deafness. Autosomal recessive inheritance was suspected because her parents were first cousins. She had mild to moderate intellectual disability. Craniofacial examination showed the presence of marked ocular hypertelorism, telecanthus, large anterior fontanelle, wide metopic suture, widow's peak in anterior hairline, depressed nasal bridge, short nose, and posterior rotation of the ears (low set ears). The facial appearance, although not coarse, was characteristic. Other findings were mid-face hypoplasia a broad forehead, exotropia, frontal bossing and wide anterior fontanel, downslanting palpebral fissures, short nose with a broad tip, cubitis valgus and posterior rotated ears and webbed neck (Figure 2). Ocular findings included enlarged globes (leading to the appearance of prominent eyes) and downslanting palpebral fissures. Ocular measurements were 45 for inner canthal and 65 mm for interpupillary distances. Intraocular pressure in right eye was 12 mmHg and left eye was 19 mmHg. Left exotrophia of approximately 45º was seen. She had strabismus. She did not have keratokonus, myopia, retinal distrophia, coloboma or cataracts. Omphalocele (or umbilical hernia) was present. Sensorineural hearing loss was positive as mesured by audiometric testing. Neuroradiologic eaxmination showed mucosal thickening in the right maxillary sinus and ethmoid cells. Odontoid process and clivus were posteriorly dislocated towards foremen magnum and compression and edema was observed at the level of C1 spinal cord level. At the level of C2, expansion of the central channel was seen. At this level, the spinal canal was narrowed, in accordance with cervical spinal cord atrophy (antero-posterior diameter of six mm). MRI scan of the brain confirmed agenesis of corpus callosum and presence of arachnoid cyst (2 cm). The cervical spinal stenosis (6 mm) was thought to be secondary to hydrocephalus. Developmental delay was present with short stature (height - 144 cm, weight - 47.5 kg) Karyotype determined on white blood cells were normal (46, XX), (Figure 3). Full abdominal ultrasound (USG) scan showed normal liver contours, size and parenchymal echogenicity. Intrahepatic and extrahepatic biliary tract, portal vein, hepatic veins and porta hepatis was normal. USG scan showed large number of stones in the gallbladder measuring few mm size and right ovarian cyst measuring 26x17 mm in size. Spleen size was 125 mm and contours and parenchymal echogenicity were normal. Pancreatic size, parenchymal thickness and echogenicity were normal. Both kidneys were normal in

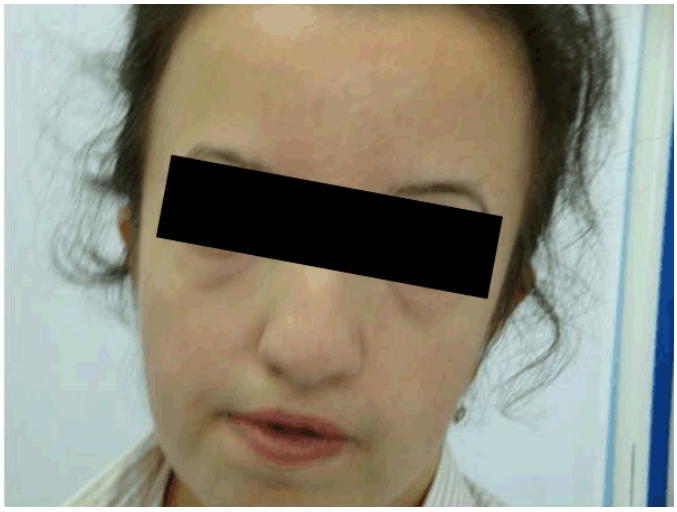

Figure 1: Facial dysmorphism is including a severe hypertelorism with downslanting palpebral fissures, a short bulbous nose, and posteriorly rotated ears were present.

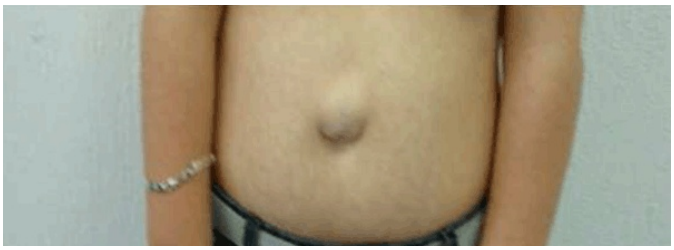

Figure 2: Umblical henia in the patient.

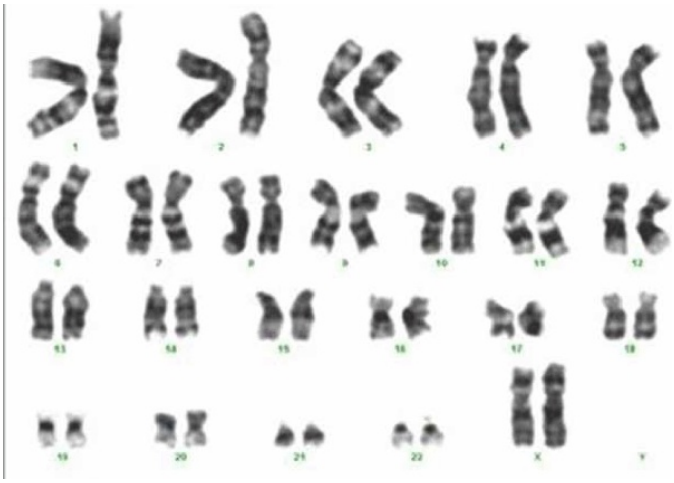

Figure 3: Cytogenetic analysis demonstrated normal karyotype (46, XX).

location and dimensions (right kidney - 91x36 mm, parenchymal thickness of 10 mm; left kidney - 97x34 mm, parenchymal thickness 11 mm). Renal contours, parenchymal echogenicity and thickness were normal. The bladder contours, the wall thickness and the lumen were normal. Left ovary was normal. In the lobe of the

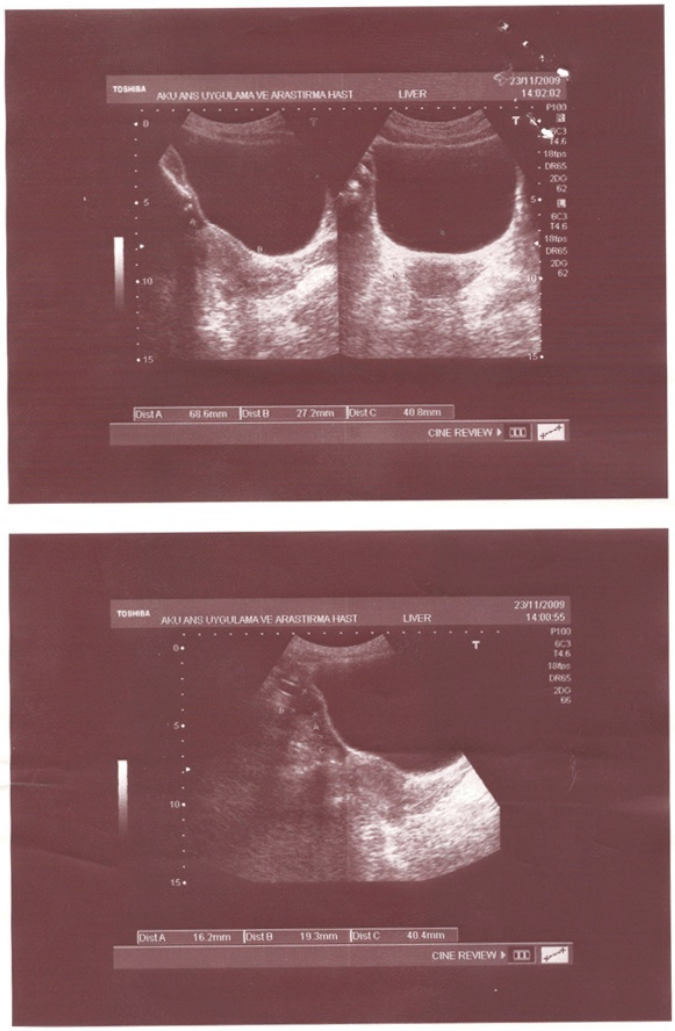

Figure 4: Pelvic ultrasound scan of uterus and ovaries.

right ovary there was a cyst measuring 26x17 mm. Pelvic USG showed the largest size follicle measuring 16 mm size in the right ovary and was otherwise normal (Figure 4).

DISCUSSION

Donnai–Barrow syndrome has an autosomal recessive inheritance pattern. In our patient's the mother and father were first-degree relatives and the inheritance pattern was autosomal recessive. Other studies also report an autosomal ressive pattern of inheritance [1, 6–8]. The patient was referred to our clinic with a presumed diagnosis of Turner syndrome. Cytogenetic analysis to assess the metaphase chromosomes of peripheral blood lymphocytes showed 46, XX karyotype in our patient. This findings in our patients were very similar to those described in this syndrome. [8]

For Donnai–Barrow syndrome, Pober et al. gave a classification in 2009. According to this classification; Donnai–Barrow syndrome and FOAR syndrome, referred to as DB/FOAR syndrome, is a unique malformation complex. Based on this review of published cases the core features of this syndrome consist of:

1. Congenital anomalies found in approximately 90% patients: hypertelorism; partial or complete agenesis of the corpus callosum; enlarged anterior fontanelle; characteristic facial features.
2. Functional anomalies found in approximately 90% patients: proteinuria; high myopia, sensorineural hearing loss; developmental delay.
3. Anomalies found in approximately 50% patients: congenital diaphragmatic hernia and omphalocele/umbilical hernia; additional features such as coloboma and macrocephaly also appear to occur in approximately 50% of cases, but the numbers are too small to state this with certainty [9].

Some of the major malformations were also seen in our patient such as; hypertelorism, telecanthus, hydrocephalus, enlarged anterior fontanelle, characteristic facial features, proteinuria, starbismus, sensorineural hearing loss and developmental delay. Diapdhragmatic hernia, as in umbilical hernia, has been observed in our patient's.

CONCLUSION

Donnai–Barrow syndrome is an autosomal recessive disease. Short stature, characteristic facial appearance, hearing loss and umbilical hernia point towards a diagnosis of Donnai–Barrow syndrome. Although a large number of malformations are present, a normal karyotype makes Donnai–Barrow syndrome an extremely interesting entity. Moderate levels of mental life is consistant with the disease and patients easily adapt to social life.

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Author Contributions

Serap Tutgun Onrat – Substantial contributions to conception and design, acquisition of data, Cytogenetical Analysis and interpretation of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published
Yaşar Sıvaci – Substantial contributions to conception and design, acquisition of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published
Ersel Onrat – Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published
Tülay Köken – Acquisition Biochemical data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CLINICAL IMAGES OPEN ACCESS

Adrenal fusion anomaly

Sunil Bylappa Kumar, Prashant Basavaraj Mahalingashetti, Ramaswamy Anikode Subramanian, Suneeta Padhy

CASE REPORT

A 35-year-old female, G2P1L1, underwent an antenatal scan at 36 weeks. Large thoraco-lumbar neural tube defect and Arnold-Chiari malformation type II were detected in the fetus. A female fetus weighing two kg was delivered following induction for termination of pregnancy. Autopsy confirmed Arnold-Chiari malformation, and meningomyelocele at thoraco-lumbar region. Histology showed features suggestive of amniotic fluid aspiration in non-aerated lungs. A transverse band of tissue across the midline was noted above the kidneys, posterior to aorta. No separate adrenals were identified in superior poles of kidney (Figure 1A, B). Histologic examination of transverse band of tissue showed a capsulated tissue with cells displayed in zonal pattern. A compact cellular layer formed outer layer, columns of paler cells in the middle and smaller cells in cords on inner side consistent with adrenal gland morphology (Figure 2). This confirmed the diagnosis of adrenal fusion anomaly.

Figure 1: (A) Abnormal midline band of tissue in the retroperitoneum (Gross picture - view from posterior aspect), (B) Closer view.

DISCUSSION

Adrenal gland agenesis, hypoplasia and accessory gland are common congenital anomalies of adrenal glands. While fusion anomalies of kidney are relatively common (1 in 300 pediatric autopsies), congenital fusion of adrenal glands is a rare anomaly with only 65 cases reported in literature [1, 2]. Most of them are detected at autopsy [1] and few as an incidental finding in imaging studies associated with other anomalies [1, 2]. These anomalies are functionally normal [2]. The fusion occurs across the midline, with resultant horseshoe or butterfly shaped gland. Fusion can be preaortic or postaortic [1, 2]. Normal histology of fused glands implies that the defect in embryogenesis occurs probably at 5–7th week. Similar case reports confirm the constant association with midline central nervous system defects such as meningomyelocele and Arnold-Chiari malformation. This confers the postulation that adrenal glands develop from single primordial gland rather than separate right and left glands. Horseshoe adrenal is less common component of asplenia

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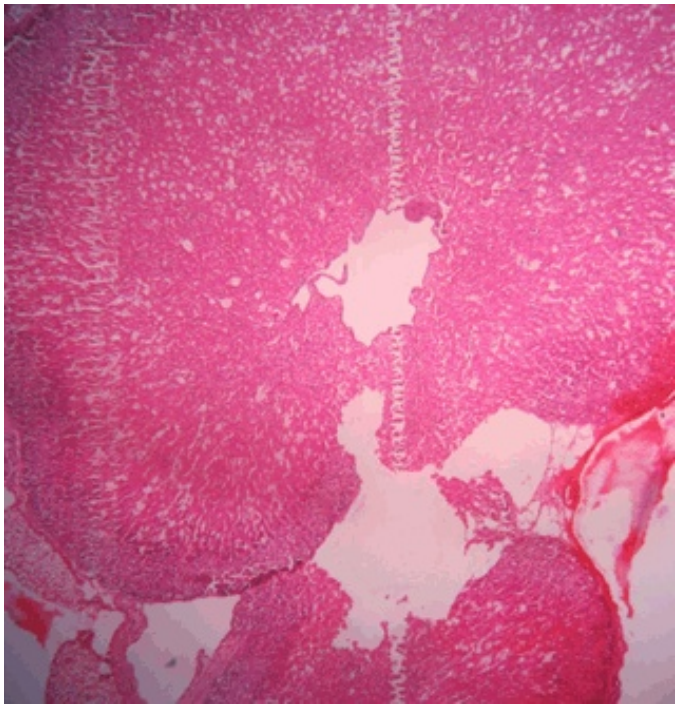

Figure 2: Cells arranged in three histological zones beneath the capsule (H&E, x100).

syndrome, a form of heterotaxy characterized by bilateral right-sidedness. Adrenal fusion may be considered as a differentiating feature between asplenia and polysplenia [3]. The management of such cases depends on type and severity of associated anomalies, their response to surgical intervention [2] and was detected antenatally which ends in termination as in our case. The occurrence of this anomaly in successive pregnancies is not mentioned in literature.

CONCLUSION

Congenital fusion of adrenal gland can be an incidental radiological finding. Awareness of this rare entity can avoid unnecessary interventions since they are histologically and functionally normal and no intervention is required but screening for potential assosciated anomalies such as central nervous system malformations, renal agenesis, asplenia, anomalies of the internal genitalia and complex cardiac anomalies are warranted.

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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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CLINICAL IMAGES OPEN ACCESS

A simple radiological solution for a neurological dilemma

Kavitha B, Balasubramanian R

CASE REPORT

A 55-year-old male presented with repeated right focal seizures of 15 days duration (3–4 episodes per day, each episode lasting for 2–3 minutes).There was no associated headache, fever or neck stiffness. There was no past history of diabetes or hypertension. He was not addicted to tobacco or alcohol and was not a pork eater. Clinically, he was well built, nourished and afebrile. There was no pallor or lymphadenopathy, and his pulse and blood pressure were within normal limits. His neurological examination was unremarkable. His blood count was remarkable for 16% eosinophils and his serum chemistry was normal. His chest X-ray was normal. Magnetic resonance imaging (MRI) brain (T1 sequence) showed a solitary left cortical ring enhancing lesion, with no definite scolex visible within it (Figure 1). Hence a possibility of neurocysticercosis (NCC) though strongly suspected, could not be confirmed and an alternative and more common possibility of tuberculoma also could not be ruled out. A serum ELISA test to demonstrate antibodies to cysticerci would not have been helpful as the lesions were not numerous. At this juncture an X-ray of the thigh (Figure 2) yielded the much needed proof in the form of the typical 'rice grain' or 'cigar-shaped' calcified cysticerci embedded in the thigh muscles. The patient was accordingly treated with antihelminthics (albendazole 400 mg b.d. for 14 days) along with intravenous dexamethasone 4 mg t.d.s. and antiepileptics (carbamazepine 200 mg t.d.s. which was continued) with a good outcome.

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DISCUSSION

Cysticerci are the larval forms of the tapeworm *Taenia solium*, residing in the human tissues like the skeletal muscle, brain, CSF, subcutaneous tissue or eye. *Neurocysticercosis* is a common parasitic disease of the central nervous system. It poses a diagnostic challenge, especially in a tropical country like India where tuberculoma can have a similar presentation and radiological features like NCC. We presented a case with one such experience.

Seizure (either focal or jacksonian) is the most common presenting symptom of NCC (70–90%). NCC is

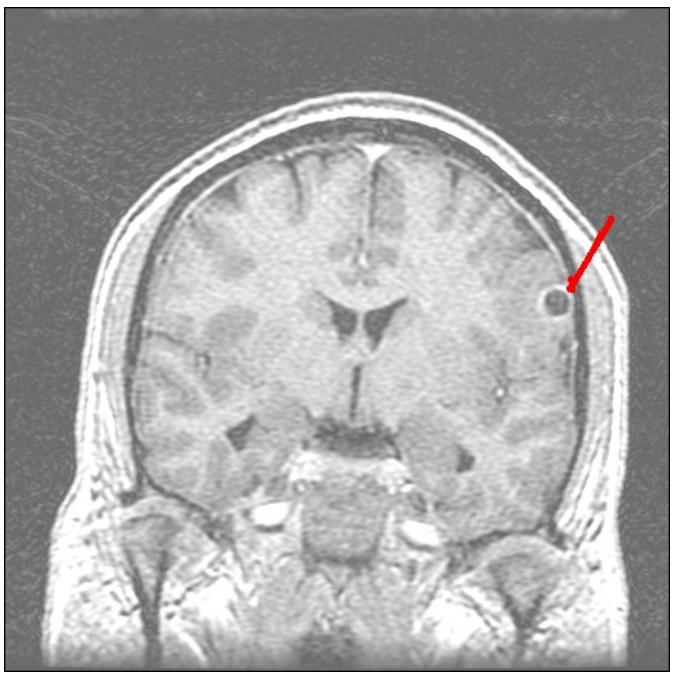

Figure 1: MRI of brain (T1 sequence) with contrast showing a solitary left cortical ring enhancing lesion, with no definite scolex visible within it indicated by the red line.

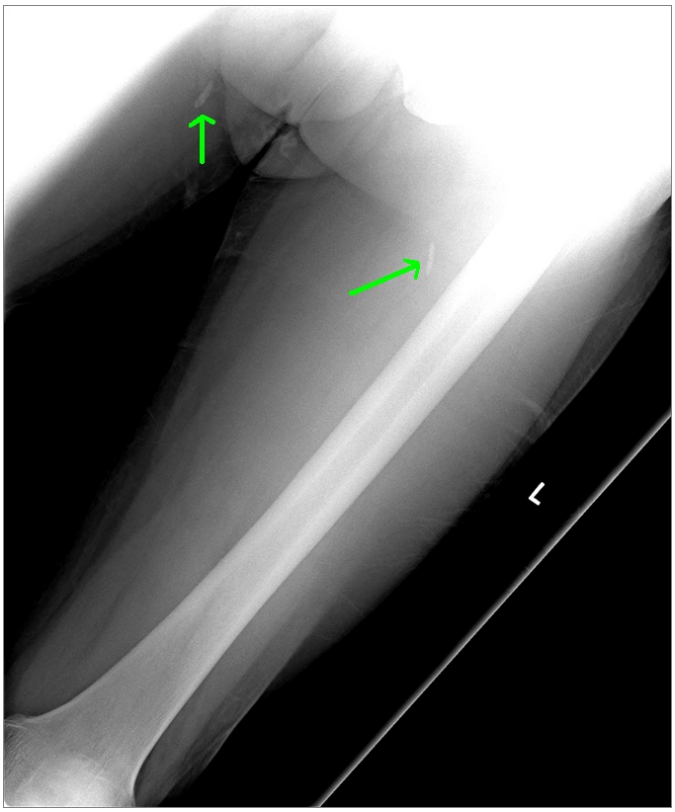

Figure 2: X-ray of the thigh showing the typical 'rice grain' like calcified cysticerci embedded in the thigh muscles, which are indicated by the green lines.

the main cause of acquired seizures in developing countries. Features of obstructive hydrocephalus due to CSF flow obstruction by cysticerci may be present.

Diagnostic Dilemma: The diagnosis of NCC can be difficult. A consensus criteria has been put forth by Brutto et al. for diagnosis [1], but, the majority of Indian patients with NCC cannot satisfy several items even of these diagnostic criteria. Non-enhancing cystic lesions on computed tomography (CT) or magnetic resonance (MR) imaging showing scolex are seen in only a small fraction of patients with NCC. In India, the overwhelming majority of patients of NCC have single enhancing lesion; multiple enhancing CT/MR lesions are also uncommon [2]. These single or multiple lesions pose a challenge to both radiologists and clinicians to differentiate it from tuberculoma, which has very similar clinical and imaging features as NCC [3, 4]. This distinction is an important issue because of the contrasting treatment for either lesion.

On MRI, the cysticercus granuloma shows a ring pattern of enhancement after contrast administration. Usually, the lesions are <20 mm in diameter. Calcified eccentric scolex is often seen in a cysticercal lesion. The lesions are often multiple and most often do not have extensive edema [5, 6]. Rajshekhar et al. noted that cysticerci are usually round in shape, 20 mm or less in size with ring enhancement or visible scolex. Cerebral edema severe enough to produce midline shift or focal neurological deficit is not seen often. Tuberculomas are usually irregular, solid and greater than 20 mm in size. They are often associated with severe perifocal edema and focal neurological deficit [8]. In India, solitary cysticercus granuloma account for 60% of cases of neurocysticercosis [7].

Positive serum EITB assay for the detection of antibodies against *T. solium* has a specificity approaching 100% and a sensitivity of 94–98% for patients with two or more lesions and 60–85% for patients with a single lesion [8]. A major weakness of this test is frequent false negative results in patients with a single intracranial cysticercus lesion (50%) and in calcified lesions [9, 10]. Access to Enzyme-linked immunoelectrotransfer blot (EITB) in India is limited. Our patient had a single ring enhancing lesion and hence we did not consider a serological test, but went for a much simpler and readily available investigation which helped us resolve the doubt. X-ray of the soft tissue shows multiple elongated foci of calcification just about the shape and size of grains of rice. These 'rice grain' calcifications are usually oriented along the direction of the muscle fibers [11]. In some series, calcified larvae have been demonstrated in up to 97% patients examined five or more years after the infection; such a high rate of detection is not to be expected routinely. Some patients with cerebral cysticercosis will have no evidence of calcified cysts in the muscles and are unaware of their infection.

CONCLUSION

NCC is a commonly encountered parasitic disease in India and worldwide. It poses a diagnostic problem for physicians as it has similar clinical and radiological features of a tuberculoma which is much more common in India. A single ring enhancing lesion could create a dead end especially when serological tests are negative too. In such circumstances, X-rays of the soft tissues like the calf/thigh muscles could reveal the calcified 'rice-grain' like cysticerci along the muscle fibers confirming the diagnosis of NCC.

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LETTER TO EDITORS OPEN ACCESS

Linezolid induced lactic acidosis and hepatic failure

Evangelos Potolidis, Eleftheria Gousi, Dimitrios Delios, Eleftheria Bei, Charalampos Mandros

To the Editors,

Linezolid is oxazolidinone antibiotic, widely used to treat infections from multidrug-resistant gram positive microorganisms by compromising bacterial ribosome function [1]. Gastrointestinal disturbances, myelosuppression, serotonin syndrome, optic neuropathy and lactic acidosis comprise the constellation of the well known linezolid associated adverse effects [2, 3]. Given the similarities between bacterial and mitochondrial ribosome, this linezolid-induced lactic acidosis is well expected [4]. This potentially serious side effect occurred mainly during long-term treatment, as depicted from the maximum four-week treatment FDA approval. We describe a patient who developed lactic acidosis and hepatic failure during a few days period of linezolid administration.

A 70-year-old female was admitted to the hospital due to fever and hypotension. The patient had a past history of hypertension and osteoarthritis. On evaluation she appeared tired and restless. The blood pressure was 90/65 mmHg, pulse 124 beats per minute, temperature 37.9°C, respiratory rate 28 breaths per minute and Glasgow coma scale 15/15. The abdomen was distended with normal bowel sounds, cardiac sounds and lungs were clear and all other examinations were normal. Chest X-ray was also normal and computed tomography (CT) scan of the abdomen was ordered due to abdominal findings and concomitant hypotension. Abdominal CT scan was normal and rapid test for influenza was negative. Specimens of blood and urine were sent for culture and cerebral fluid was drawn. Cerebral fluid was negative. The blood tests revealed elevated WBC to be $15x10^3/mm^3$ and increased C-reactive protein levels. Other routine tests were normal including liver enzymes. Soon after, the patient became hemodynamically unstable. Therefore linezolid was administered (600 mg b.d.) and ceftazidim (2 g t.d.s.). Fluids and vasopressors were given and the patient was temporarily admitted to the intensive care unit. Urine culture revealed urinary tract infection due to pseudomonas. Blood cultures were negative. Cardiac ultrasound was normal. Chest X-ray was performed on the second day because of breathing discomfort and revealed a left upper lobe pneumonia. CT scan of chest confirmed the inflammatory process of the left upper lung lobe. On the third day, the patient was afebrile, hemodynamically stable with normal urine output. On the sixth day, the patient suffered from abdominal discomfort. On examination, the patient was alert but appeared uncomfortable. Vital signs were normal. Abdomen X-ray and ultrasound (USG) scan were normal. Blood tests revealed total bilirubin 2.2 mg/dL (0.3–1.2 mg/dL), indirect bilirubin 1.7 mg/dL, SGOT 234 IU/L (5–40 IU/L), SGPT 222 IU/L (5–40 IU/L), γGT 100 IU/L (10–50 IU/L), LDH 443 IU/L (130–245 IU/L) and INR 2.8 (<1.2). Further laboratory investigations depicted a high anion gap metabolic acidosis with bicarbonate 6 mEq/L (24 mEq/L), anion gap 48 mEq/L (12 mEq) (normal range in brackets), elevated lactate levels 20 mmol/L and renal failure (normal range ig given in brackets) (Cr 2.3 mg/dL (0.9–1.6 mg/dL)). Methanol was not detected. On the next day, the possibility of linezolid toxicity was considered and the drug was withdrawn. The patient was switched to clarithromycin 500 mg twice daily and ceftazidim resulting in a normalization of liver enzymes, lactate levels, prothrombin time and renal function.

Lactic acidosis is the most common cause of metabolic acidosis in hospitalized patients. Type A lactic acidosis (due to tissue hypoperfusion) is observed in septic shock, hypovolemia and cardiopulmonary arrest.

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Metformin and nucleoside reverse transcriptase inhibitors are able to cause type B lactic acidosis (mitochondrial dysfunction). As previously mentioned, linezolid has the potential to inhibit mitochondrial ribosomes. The resulting deficiency of the thirteen mitochondrial DNA encoded proteins results in mitochondrial malfunction. Interestingly this effect is observed predominately after prolonged drug exposure. Additionally, lactic acidosis is reported to be dose dependend. In patients treated with the standard 600 mg dose every 12 h, overexposure was documented in 12% cases. It has been suggested that overexposure occurs especially in patients cotreated with drugs which may act as P-glycoprotein inhibitors (omeprazole, amiodarone). The implementation of therapeutic drug monitoring may be helpful limiting such cases [5]. Patients with mitochondrial diseases such as MELAS syndrome (mitochondrial encephalomyopathy, lactic acidosis and stroke like episodes) are prone to lactic acidosis after linezolid treatment [6].

Our patient developed lactic acidosis and liver dysfunction during treatment with linezolid. Undoubtably linezolid is a valuable drug with a good safety profile. Clinicians need to be aware of linezolid-induced lactic acidosis.

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Dimitrios Delios – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Eleftheria Bei – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Charalampos Mandros – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

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Conflict of Interest

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